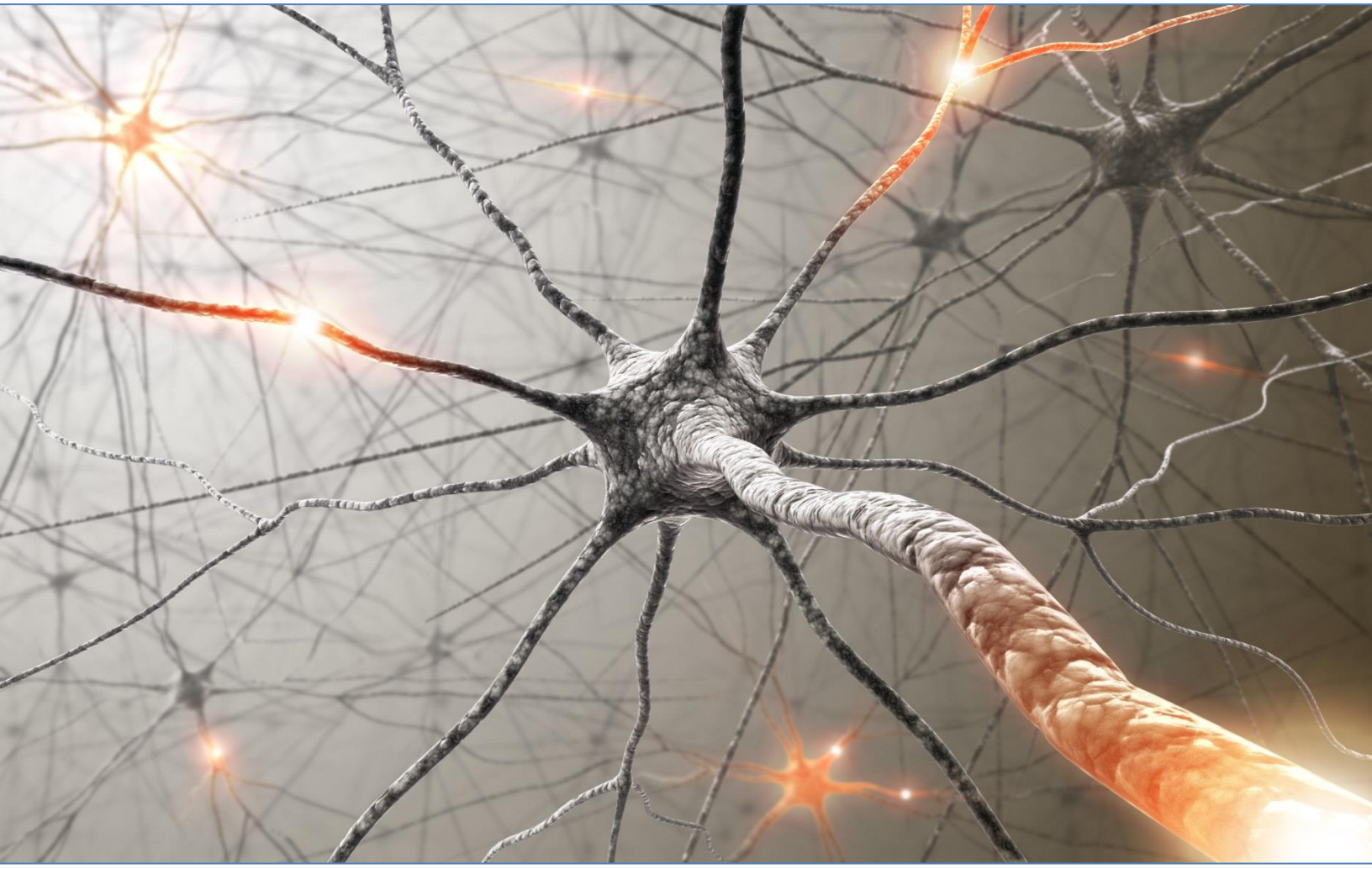
]]NEW EDITION

Paediatrics
Neurological disorders

By Dr ali bel kheir



يخاطبني السفيه بكل قبح..... فأكره أن أكون له مجيبا
يزيد سفاهة فأزيد حلماً.....كعود زاده الإحراق طيبا
إذا نطق السفيه فلا تجبه..... فخير من إجابته السكوت
فإن كلَمته فرَجت عنه .....وإن خليته كمداً يمـوت
(الامام الشافعي)

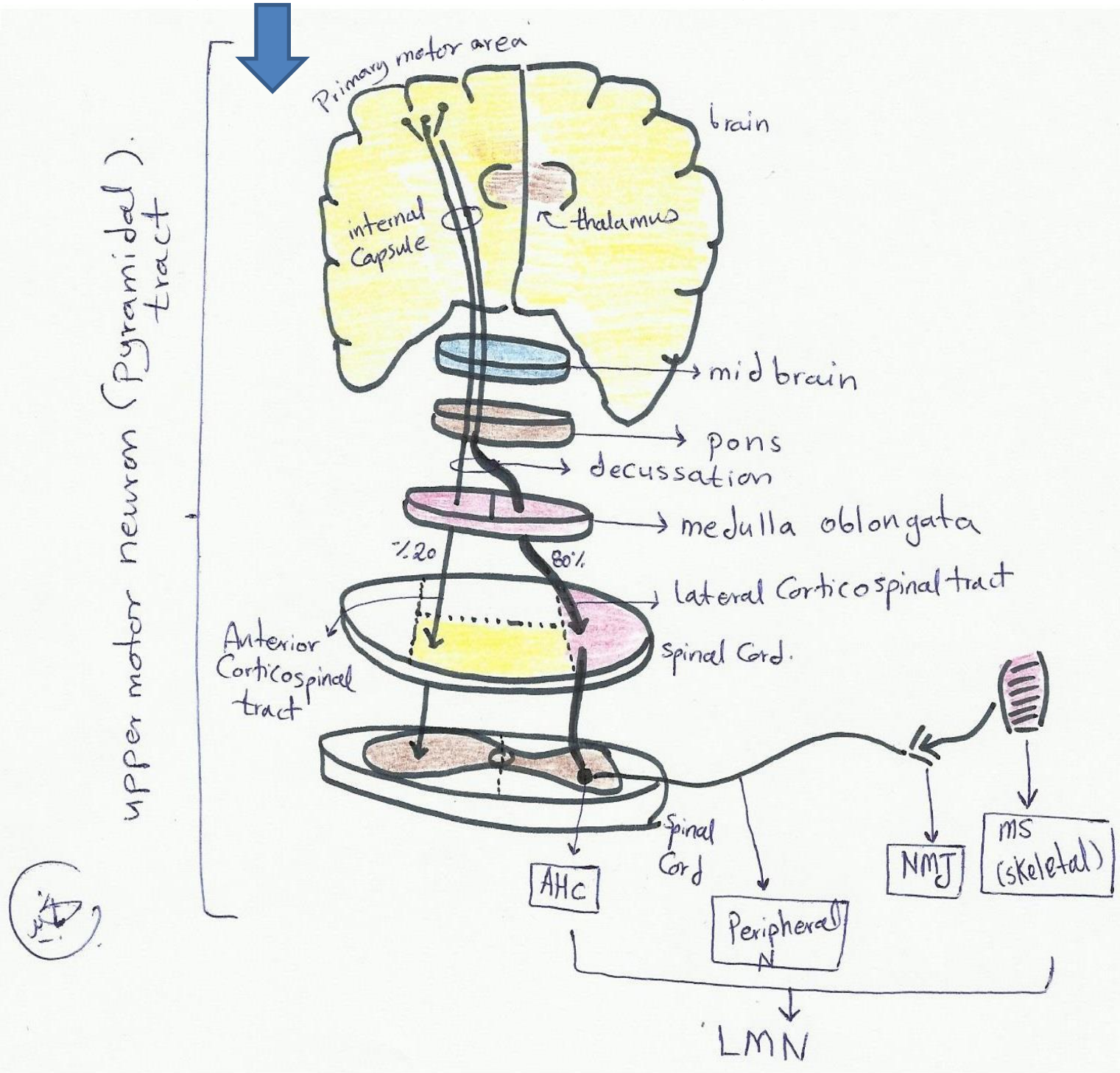
INDEX:

Motor neuron lesion

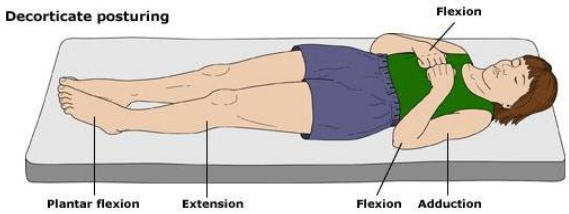
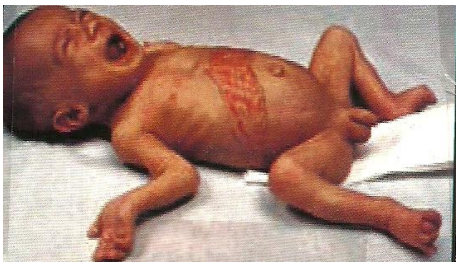
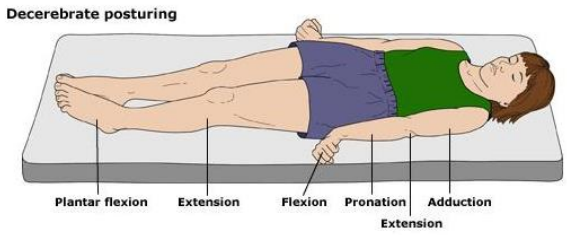
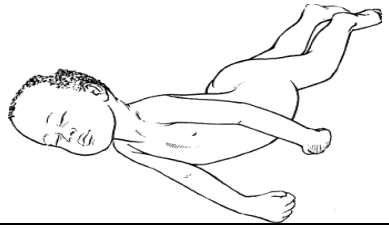
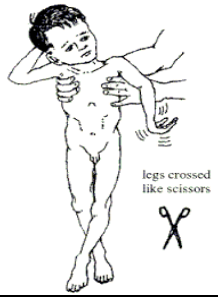
- Divided into→UMNL and LMNL
- **UMNL**→Any lesion above AHC.
- **LMNL**→any lesion from AHC to Ms

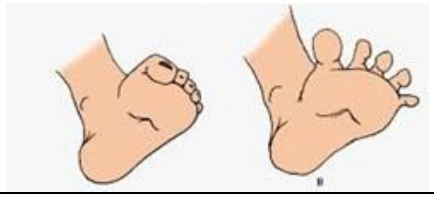
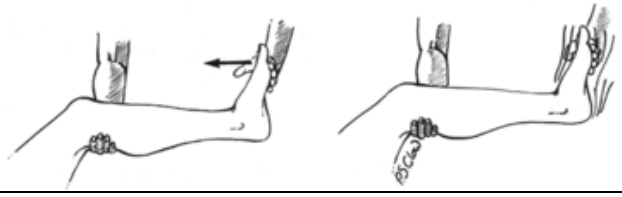
UMNL

Pyramidal lesion **extra-pyramidal lesion**



CLINICALLY YOU CAN DIFFERENTIATE:

	UMNL	LMNL
inspection *posture :*	**1.Decorticate position:**  Result from damage to one or both corticospinal tract	**1. Frog like position:** (Due to hypotonia). 
	2.Decerebrate position:  Result from damage to the upper brain stem	
	3.Frog like position: ❖ Only if baby below 1 yr (infant) or in early shock stage.	
	4.Oppostitone: -arched back and neck. 	
	5. Scissor position:  	
muscle wasting:	(-) ❖ Except in long standing.	(+)

<table>
<tr><td rowspan="2"><u>fasciculation:</u></td><td>(-)</td><td>(+)</td></tr>
<tr><td colspan="2">❖ its involuntary movement of group of ms. fibres</td></tr>
<tr><td>Palpitation:
<u>ms tone:</u></td><td colspan="2">Hypertonia
Pyramidal → (spastic) Clasp knife
extra pyramidal (rigidity)
Cog weal (Rigidity + tremor)
lead pipe (Rigidity only)</td><td>hypotonia</td></tr>
<tr><td><u>power:</u></td><td colspan="3">Grades:
0= complete paralysis.=paralysis
1= only fasciculation.
2= with gravity. From 1 to 4=paresis=ms weakness
3= against gravity.
4= with mild resistance.
5= complete resistance=normal</td></tr>
<tr><td><u>Reflex:</u></td><td colspan="2">(+)
Brisky
Hyper-reflexia</td><td>(-)
decreased</td></tr>
<tr><td rowspan="2"><u>Babinski sign:</u></td><td colspan="2">(+)</td><td>(-)</td></tr>
<tr><td colspan="3">Babinski: dorsiflexion of the big toe with fanning of others
Toes- its Normal response below 1 year
  </td></tr>
<tr><td rowspan="2"><u>Clonus:</u></td><td colspan="2">(+)</td><td>(-)</td></tr>
<tr><td colspan="3">Clonus: slightly flexion of knee joint and ankle then sudden dorsiflexion of ankle → repeated contraction of calf ms →continued stretch (normal below 2mts of age)
  </td></tr>
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DIFFERENCE BETWEEN PYRAMIDAL AND EXTRA PYRAMIDAL LESIONS:

	pyramidal	**Extra pyramidal**
tone	Clasp knife spasticity.	Lead pipe or coq wheel.
Deep reflex	Hyperreflexia	normal

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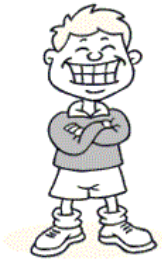
LMNL
Peripheral motor disorders: The neuromuscular disorders



- Any part of the lower motor pathway can be affected in a neuromuscular disorder: هام

AHC lesion	1. Werdnig Hoffmann disease. 2. Poliomyelitis.	Signs of denervation(motor)= weakness, loss of reflexes, fasciculation and wasting
Peripheral nerve lesion	1. Hereditary motor sensory neuropathies 2. Acute post-infectious polyneuropathy (Guillain-Barré) 3. Bell's Palsy	(Motor=weakness)and sensory impairment
NMJ lesion	1. Myasthenia gravis. 2. Botulism	Fatigability Motor+sensory
Ms lesions	**1. Muscle dystrophies.** - Duchenne/Becker/ congenital **2. Inflammatory myopathies.** - Benign acute myositis - Polymyositis/ dermatomyositis **3. Myotonic disorders.** - Dystrophia myotonica **4. Metabolic myopathies.** **5. Congenital myopathies.**	Weakness→often proximal Wasting, gait disturbance. May be with sensory involvement

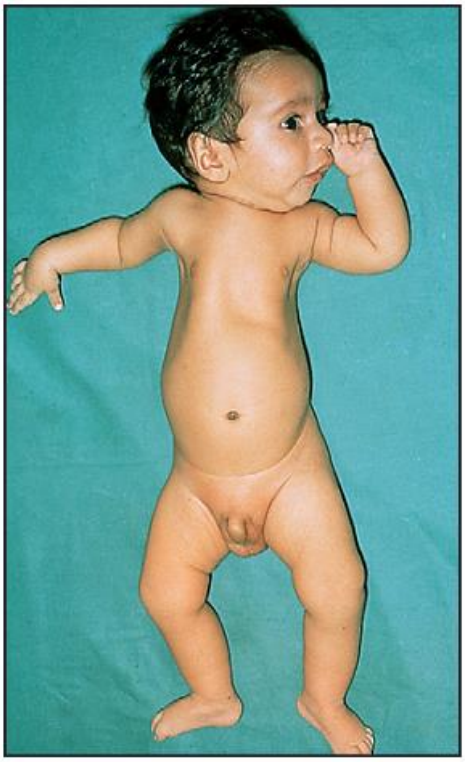
Disorders of the anterior horn cell
Spinal muscular atrophy type 1
(Werdnig–Hoffmann disease)

- Autosomal Recessive.
- One of the commonest causes of floppy infant
- This is the second most common cause of neuromuscular disease in the UK after Duchenne muscular dystrophy
- Very severe progressive disorder presenting in early infancy

PATHOPHYSIOLOGY:

- AR→mutations in the survival motor neurons (SMN) gene→progressive atrophy of AHC (motor neurons)in spinal cord→weakness and wasting of skeletal muscles

CLINICAL PICTURES:



- Onset before 2 years of life (often start in uterus)
- ↓fetal movement during pregnancy
- Arthrogryposis at birth
- Generalized ms weakness.
- Lack of antigravity power in hip flexors
- Absent deep tendon reflexes
- Intercostal recession
- Floppy infant → frog like position.
- Sever hypotonia of proximal and distal limps.
- Tendon reflexes are absent.
- Infant with normal intelligence.
- Fasciculation is visible mainly in tongue.

INVESTIGATION:

1- **CPK** = creatinine phosphokinase (normally or slightly high).

2- **EMG** = neuropathic→fibrillation and evidence of ms denervation.

3- **Nerve conducting study**= slow conduction and denervation study.

4- **Ms biopsy**=denervation of ms (ms is not involved, it`s important to Differentiate it from congenital myopathy).

TREATMENT:

1- No medical treatment.
2- Only supportive.

PROGNOSIS:

- Most of pts die before 2 years (mainly 12 mts) secondary to respiratory failure and food aspiration.

NOTES:

- There are milder forms of the disorder with a later onset Children with type 2 spinal muscular atrophy can sit, but never walk independently.
- Those with type 3 (Kugelberg Welander) do walk and can present later in life.

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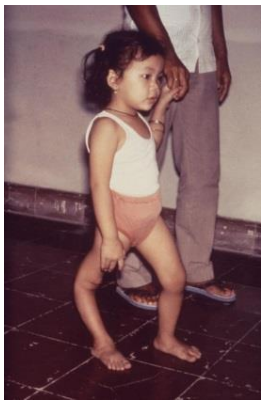
Poliomyelitis



CAUSED BY→ polio virus→small RNA viruses (picorna virus)

SPREAD BY→fecal –oral route and air droplets

PATHOPHYSIOLOGY→destroy AHC→AFP

CLINICAL PICTURES:

AFP-Asymmetrical-ascending ms weakness

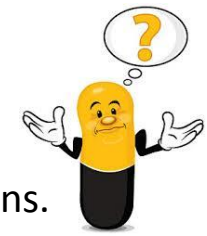
Nausea, vomiting,abdominal cramps,pain,diarrhea,sore throat for 1-3 days

Fever, neck stiffness→aseptic meningitis

Asymmetrical ascending ms weakness→AFP→permanent weakness

INVESTIGATION:

- **Diagnosis done clinically and by isolation of virus** from stool sample or a swab of the pharynx→ Antibodies to poliovirus can be diagnostic
- **PCR** to determine the source of the virus
- **Csf**→aseptic meningitis (↑WBC (lymphocyte)-↑protein)



TREATMENT:

- There is no cure for polio→symptomatic and preventing complications. Ex: antibiotics for infection
- Vaccination→WHO 1988-1994 eradicate the polio in developed countries
- But Some cases still seen in Africa and immigrants

NOTES:

ACUTE FLACCID PARALYSIS

- ❖ Characterized by weakness or paralysis and reduced muscle tone without other obvious cause.
- ❖ This condition can become fatal if it affects the respiratory muscles
- ❖ Caused by:

1. Poliomyelitis
2. Guillain barre syndrome
3. Botulism
4. Myositis
5. Myasthenia gravis
6. VZ virus
7. Diphtheria
8. Rabies
9. Tick bite paralysis

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The 'floppy infant'

- ❖ Persisting hypotonia in infants.
- ❖ **Detected clinically by:**

1. frog like position
2. Ventral suspension→c shape
3. Pull to sit→head lag
4. Acrobatic sign→+
5. Scarf Sign→+
6. Slipping down

☒ **In central cuases**:

- truncal tone↓
- limbs tone preserved

☒ **Genatic causes:**

- Atonic CP

☒ **LMNL:**

- Frog like position
- Reflex ↓

Central

Cortical

- Hypoxic-ischaemic encephalopathy
- Cortical malformations

Genetic

- Down syndrome
- Prader–Willi syndrome

Metabolic

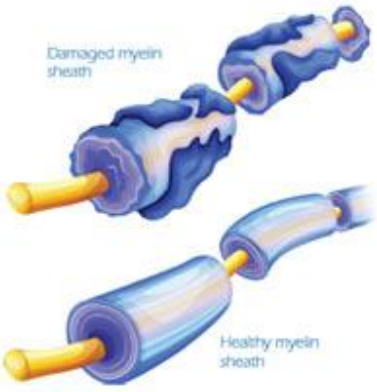
- Hypothyroidism
- Hypocalcaemia

Peripheral

Neuromuscular

- Spinal muscular atrophy
- Myopathy
- Myotonia
- Congenital myasthenia.

Peripheral neuropathies
Guillain-Barre Syndrome



Other names:

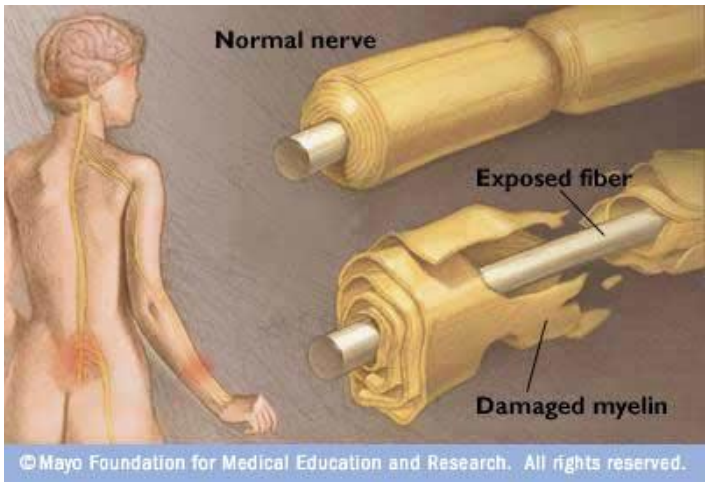
☒ Acute inflammatory poly neuropathy.
☒ Acute demyelinating poly neuropathy.
☒ Post infectious poly neuropathy.

PATHOPHYSIOLOGY:

☒ Post infection (usually viral)→ antibody attaching myelin→ Demyelination in motor, sometimes sensory nerves, Autonomic involvement

RISK FACTORS:

1. EBV.
2. Coxsackie virus.
3. Echovirus.
4. Influenza.
5. CMV.
6. Mycoplasma pneumonia.
7. Campylobacter gastroenteritis



CLINICAL MANIFESTATION: ((Most common cause of acute paralysis))

(With history)
Vaccine (influenza) (rare).
Viral illness (upper respiratory tract infection).
Campylobacter gastro-enteritis.
↓After 2-3 weeks↓

☒ ***Ascending symmetrical ms weakness***, loss of reflex on examination
☒ Autonomic involvement.
☒ Sensory symptoms usually in L.L.
☒ Involvement of bulbar ms → difficult chewing, swallowing → aspiration.
☒ 20% have bladder dysfunction.
☒ 50% have cranial nerve involvement.

(Maximum ms weakness may occur only 2-4 weeks after onset of illness)

↓↓

95% of cases will have full recovery
This may take up to 2 years

☒ ON EXAMINATION:

1. Symmetrical ms weakness/pain.
2. areflexia.
3. Little sensory involve.

☒ INVESTIGATIONS:

1- **L.P** → cyto-albumin dissociation → not seen until the 2nd week of illness-(high CSF protein) → normal cells
→ normal sugar

2-**Nerve conduction velocities** → reduced

3-**EMG** →

a) ↓Motor nerve conduction.
b) Sensory nerve conduction slow.

TREATMENT: "Supportive":

☒ Admission + monitor vital signs +o2 saturation

☒ If pt:

unstable:	stable:
Clinical indicators for intubation in the ED include the following: 1. Hypoxia 2. Rapidly declining respiratory function 3. Poor or weak cough 4. Suspected aspiration 5. Respiratory ms involved. 6. Vital capacity (FVC) is less than ↓15 ml/kg	No previous finding 
Intubate and mechanical ventilation	IV immunoglobulin, to improve rate of recovery or plasma exchange

Corticosteroid have no beneficial and may delay recovery

PROGNOSIS:

1. 95% of cases will have full recovery-This may take up to 2 years
2. 35% with permanent neurological deficit.
3. 5% may die.

The hereditary motor sensory neuropathies (HMSN)

- Group of disorder →symmetrical slowly progressive ms wasting (distal>proximal)
- Type 1 known as peroneal muscular atrophy (charcot marie tooth disease)
- Its dominant inherted
- Nerve biopsy→onion bulb formation
- Onset →first decade →distal atrophy+ pes cavus (legs>arms)
- Sensory involvement may occurs (rare)
- Its Chronic disorder (rare that pts loose ability to walk)

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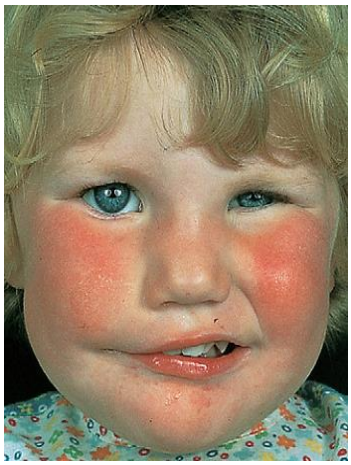
Bell palsy and facial nerve palsies

- **Bell palsy** is an isolated LMN paresis of the VIIth cranial nerve→ facial weakness



- **Causes**→unclear
  1. post-infectious→herpes simplex virus in adults.
  2. May be associated with COA , sarcoidosis , Lyme disease.
- **Presented clinically:**

1. Wrinkles on forehead absent
2. Child can't close eye forcefully
3. Cheeks puffed out→balloon on side moreThan the other side
4. Showing teeth angle of mouth is asymmetrical

- **Treated:** usually with corticosteroid→↓edema in facial canal during 1th wk

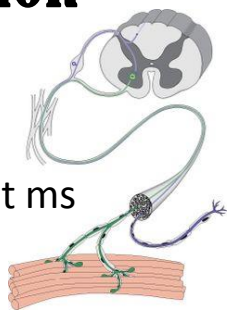
No benefit from acyclovir (except if symptoms of VIII nerver paresis Present →lesion in cerebellopontine angle)

- Complete recovery occur in majority of cases → several mts
- **complication** is conjunctival infection due to incomplete eye closer on blinking→need patch or even tarsorrhaphy
- Hypertension must be excluded (high association with COA)

Note:

☒ In superior nuclear facial palsy→superior part of face escape palsy
☒ In infra nuclear facial palsy→whole side of face is paralysed

Disorders of neuromuscular transmission
Juvenile Myasthenia gravis



DEFINITION:

☒ Autoimmune disease in which pts failure to sustain repeat straight ms contraction (fatigability).

PATH PHYSIOLOGY:

Thymus-lymphoid organs

↓

Produce β lymphocyte

↓

Antibody against acetyl choline receptor in NMJ

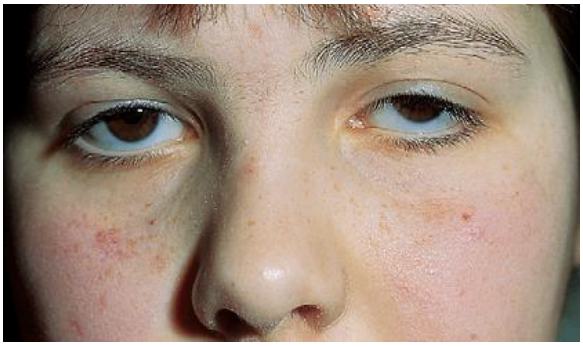
SEX:

Female > male in young

ONSET:

After 10 year old of age (late).

C/P:



1. Ptosis → extra ocular ms are first involved.
2. Loss of facial expressions.
3. Difficult of chewing.
4. Muscle weakness:
5. Proximal ms > distal ms
6. Upper limp > lower limp

INVESTIGATION:

1.Tensilon test = edrophonium test:

☒ IV injection of edrophonium chloride lead to rapid relieve of symptoms, it block or ↓break down of acetyl choline by cholinesterase.

2.Serology : identify certain antibody (antibody against ach receptors).
Seen in 60-80% of cases

TREATMENT:

1. **Oral long acting anticholinestrase** ex.: neostigmine / pyridostigmine.
2. **IV Immunoglobuline**.
3. **Immunosuppressive** as azathioprine or prednisolone is of value
4. **Thymectomy** → in case of thymoma with no response to medical treatment
5. **Plasma exchange** is used for crises

Botulism

- ❖ It is a rare and potentially fatal paralytic illness caused by a toxin produced by the bacteria Clostridium botulinum.
- ❖ The disease begins with :
  - weakness
  - trouble seeing - speaking
  - feeling tired

→ Followed with weakness of the arms, chest muscles, and legs

- ❖ The disease does not usually affect consciousness or cause a fever.

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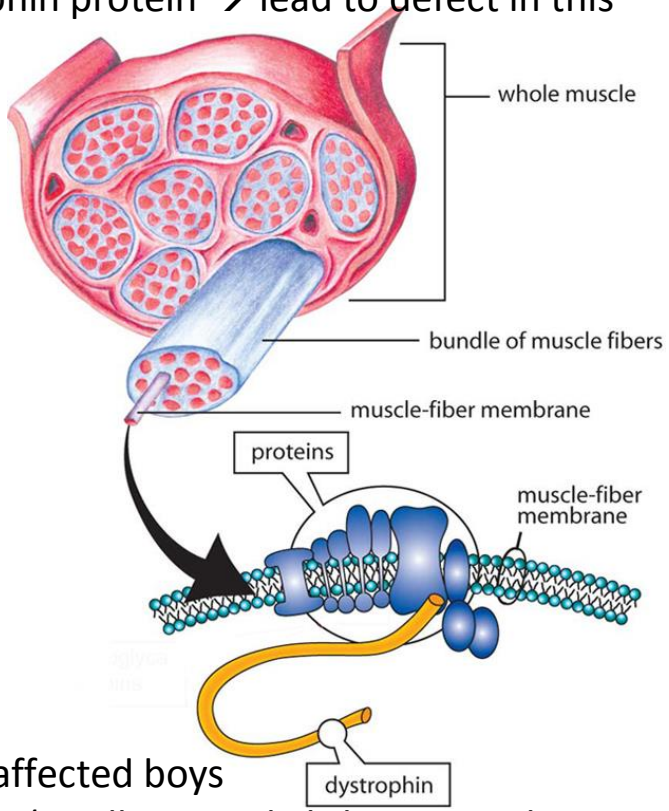
Muscle disorders
Duchennes muscular dystrophy

- ❖ Most common muscular dystrophy.
- ❖ X- Linked recessive → male affected, female carrier.

CAUSE:

- ❖ deletion of chromosome material on short arm of X-chromosome at xp-21 site, this site contain the code of dystrophin protein → lead to defect in this protein → irreversible destruction
- ❖ 30% of pts develop new mutation.



ONSET :

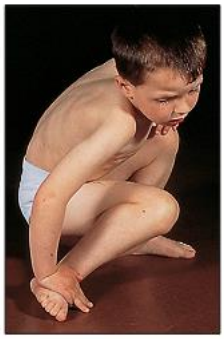
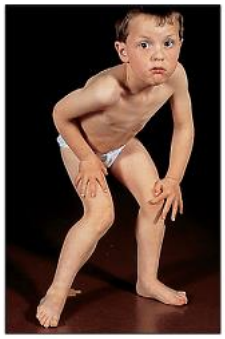
- Usually male below 5 yrs old.
-early onset.
-average age for diagnosis 5.5 years

C/P:

1. Poor head control. (1st sing)
2. Waddling gait.
3. Mount stairs one by one.
4. Learning difficulties→At school age the affected boys tend to be slower, clumsy than their peer (intelligence slightly impaired or normal but no mental retarded).

O/E:

1. Psudo-hyperatrophy of tongue and calf ms (the cause is replacement of ms fibres by fat and fibrous tissue).
2. Ms atrophy → pectoralis major
   → Bronchioradialis
3. Gower sign → seen at 3 yrs of age.
4. Skeletal deformities (including scoliosis in some cases)
5. Waddling / Trendelenburg gait → at 5-6 yrs old.
6. Wheel chair by → 12 yrs.
7. Cardiomegaly (may develop).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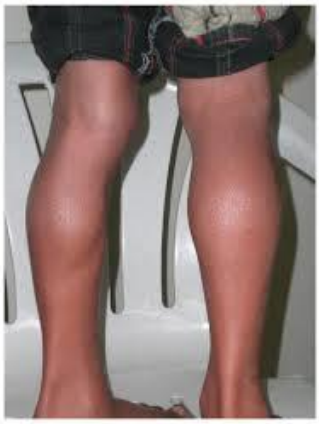


INVESTIGATIONS:

1. CPK → ↑20-200
2. EMG → myopathy changes.
3. ms-biopsy → (necrotic tissue, fat cells, fibrous tissue).

TREATMENT: " supportive "

1. Exercise→increase power strength-->
2. Physical therapy is helpful to maintain muscle strength, flexibility, and function.
3. Orthopedic appliances (such as braces and wheelchairs) may improve mobility and the ability for self-care
4. Corticosteroids such as prednisolone increase energy and strength and defer severity of some symptoms

COMPLICATIONS:

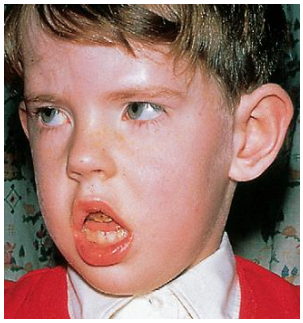
❖ Death usually at age of 18 yrs due to → respiratory failure
→ Heart failure

Backer ms dystrophy

❖ X- Linked recessive.
❖ insufficient dystrophin produced in the muscle cells
❖ Similar to Duchennes dystrophy but clinical prognosis more slowly.
❖ Symptoms usually appear in men at about ages 8–25, but may sometimes begin later.
❖ Other information similar to Duchennes muscular dystrophy

Myotonia dystrophica

- Autosomal dominant.
- Early onset myotonia = prolonged contraction, difficult to relaxing after vigorous effort due to ms fibre changes.
- caused by a nucleotide triplet repeat expansion
- affect any age



C/P:

- In neonatal period may be present with hypotonia.
- Associated with frog like position with difficult feeding and FTT.
- This manifests as slow release of handshake or difficulty releasing the tightly clasped fist-Delay eye opening after closure.
- Pts with learning difficulties.
- Associated with (cataract, testicular atrophy, diabetes, mental retardation).

TREATMENT: symptomatic

- Most of pts die due to cardiomyopathy

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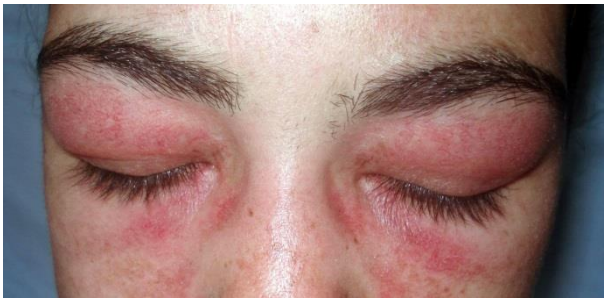
Dermatomyositis

- This is a systemic illness
- probably due to an angiopathy.
- Usual onset is between 5 and 10 years.



C/P:

1. fever, misery
2. symmetrical muscle weakness→ proximal
3. involve pharyngeal muscle→affects swallowing
4. characteristic violaceous (heliotrope) rash to the eyelids, and periorbital oedema also affect the extensor surfaces of joints, e.g. elbow, and
5. with time subcutaneous calcification can appear.

INVESTIGATION:

1. (CRP, ESR) can be raised but not invariably.
2. Muscle biopsy → inflammatory cell infiltrate and atrophy.

TREATMENT:

Physiotherapy is needed to prevent contractures.
Corticosteroids are the standard treatment, and continue at a tailored dose for 2 years. Other immunosuppressants may be needed.

Large head (macrocephaly)

DEFINITION:

❖ OFC above 97 centile according to age and sex or 2 standards Deviation above mean

CAUSES (D-D):

Cranial causes:

1. Constitutional
2. Achondroplasia
3. Familial
4. Anaemia (chronic haemolytic)
5. Rickets

Intracranial causes:

1. Hydrocephalus
2. Space occupying lesion EX. tumor
3. Subdural haematoma or effusion
4. Hydrancephaly
5. Megalencephaly which may be due to:

A. Cretinism

B. Storage Diseases
(E.X:mucopolysacharidosis)

C. Familial

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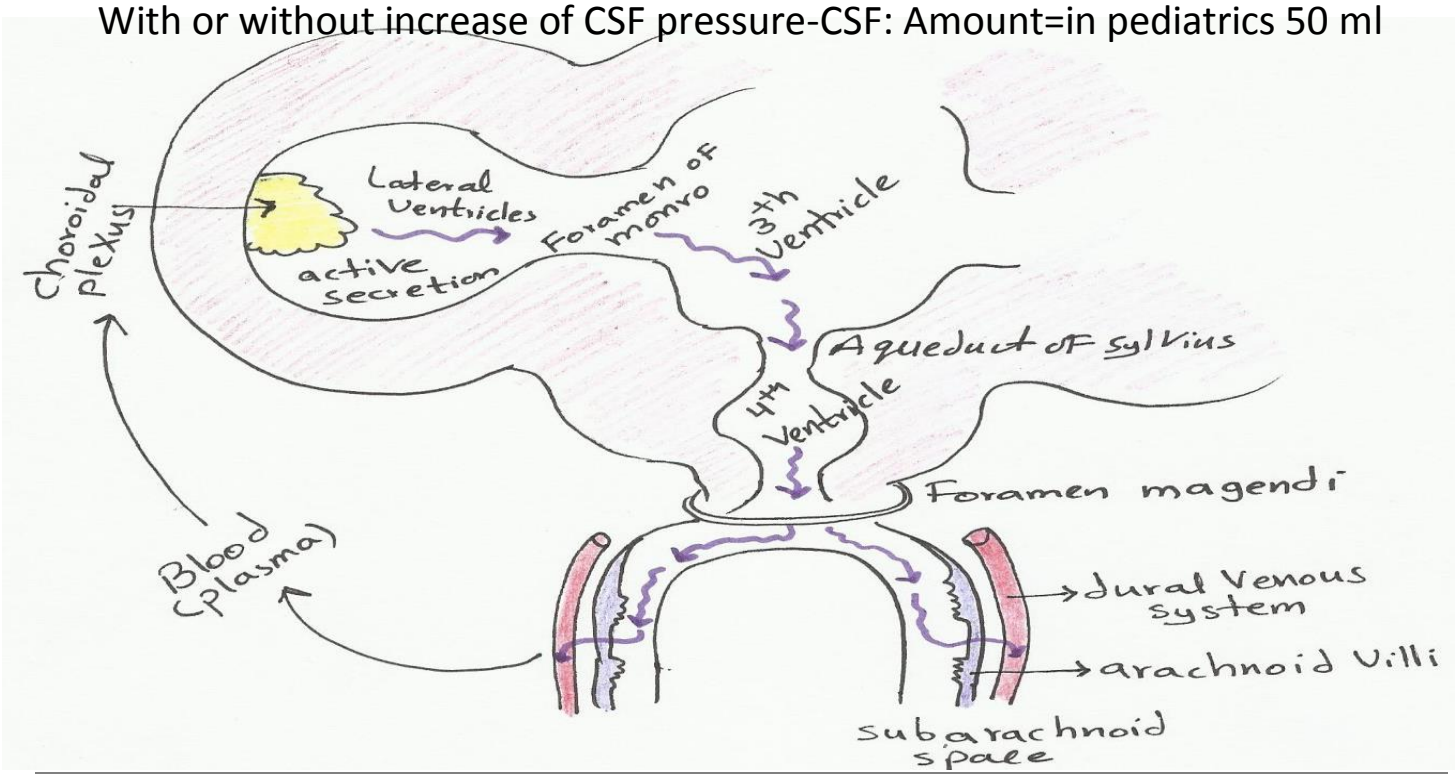
Hydrocephalus

DEFINITION:

❖ Enlargement of cerebral ventricles due excessive accumulation of CSF With or without increase of CSF pressure-CSF: Amount=in pediatrics 50 ml



CAUSES (TYPES):

Obstructive-non communicating type:

A.Obstruction of aqueduct of sylvius:

1. Congenital atresia
2. Obstruction from outside (Ex: tumers-malformation of vein of galen
3. Post Hge
4. post meningitis (TB-pneumocci-mumps)

B.Congenital atresia:

- ✓ foramen of monro
- ✓ magendi =dandy walker malformation

C.arlond chiari malformation

D.congenital infection=toxoplasmosis

E.brain tumors

Non Obstructive- communicating type:

Defect of absortion:

1. Subarachnoid space adhesion=post He-meningitis
2. Leukemic infiltration
3. Dural sinus thrombosis
4. achondroplasia

……

Excessive CsF secretion:

1. Choroidal plexus papilloma
2. Choroidal plexus congestion (as meningitis)

CAUSES OF CONGENITAL HYDROCEPHALY:

(early detection give good prognosis)

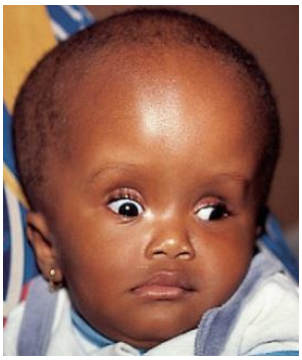
1.bleeding in fetus
2.Mother infection→toxoplasmosis-syphilis
3.Birth defect→aque duct stenosis-Spinal bifida
4.arlond chiari malformation
5.dandy walker malformation

CLINICAL PICTURES:

HEAD SIGNS:

1. Wide sutures, Fontanels are widely opened & bulging.
2. Large head with progressive increase in size (increasing head circumference on serial measurement)
3. Dilated scalp veins.
4. Eyes deviated downwards→ sunset appearance
5. Skull percussion →resonant(cracked pot sound) (**Macewen sign**).
6. Craniotabes in all bones.
7. Back of the skulls→ Promeninet occiput in= Dandy Walker.
   Foreshortened occiput in = Arnold Chiari.

8.translumination test→+ (in sever Hydrocephalus,Hydrancephaly)



NEUROLOGIC SINGS:

Mild→due fontanels and suture of skull become wide

1. Mild vomiting
2. Squint
3. Delayed motor milestones
4. Pyramidal tract lesion signs are common especially in lower limbs.
5. In advanced cases MR & optic atrophy may occur.

GENERAL EXAMINATION :

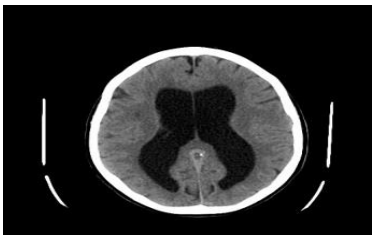
1. Back of spine for tuft of hair, lipoma or angioma in spine bifida.
2. Menjngeomyelocele in Arnold Chiari malformation.
3. Cerebellar ataxia in Dandy walker malformation.
4. Fundus examination for chorioretinitis in toxoplasmosis.

IN OLDER CHILD:

Marked neurological manifestations as the sutures are not easily separated and fontanels closed→ marked increase intracranial tension

1. Bursting headache =severe in the morning
2. Blur or vision
3. Projectile vomiting (unrelated to meals, not preceded by nausea)
4. Bradycardia & hypertension (Cushing response)

INVESTIGATIONS:



1. **Antenatal**→US
2. **CT,MRI**→Diagnostic
   Detect ventricular dilatation,cortical atrophy,and may be causes
3. **CSF**→in obstructive type→ xanthocluomia & cytoalbllminous dissociation
4. **Xray** →*before closer of sutures:* wide fontanels and sutures ,large cranium
   After closer of sutures: beaten silver appearance,wide sella

TREATMENT:

✓ Always surgical→medical treatment done before surgery

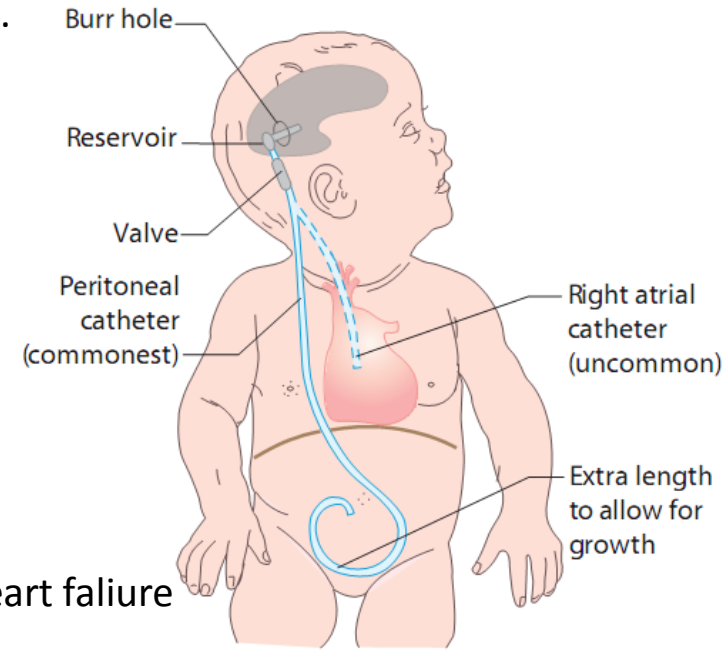
Medical:

1.Diuretics→decrease CSF→acetazolamide (Diamox tablets) or Frosemide
2.Correct Electrolyte , PH disturbances.

Surgical:

1.Extra cranial shunt operation:

1.Ventriculoperitoneal
✓ Commonest
✓ Usually on right side:
1. Left hemisphere most prominent in function
2. Heart on left side



2.Ventriculoatrial (right)→rare due: arrythmeia(SAN),infection,overload heart faliure

3.Ventriculopleural

COMPLICATION OF SHUNT:

1. Shunt nephritis (immune complex mediated)
2. Obstruction
3. Infection→staph epidermides
4. Shortness of shunt

NOTE:

❖ Shunt has ***no value*** if there:
1.Marked cortical trophy
2.MR
3.motor disability and blindness

2. Choroid plexectomy a diathermy for choroid papilloma

3.Endoscopic treatment to create a ventriculostomy

ARRESTED HYDROCEPHALUS:

- ❖ Hydrocephalus stopped on follow up
- ❖ Neurological state is stable in presence of stable ventriculomegaly
- ❖ Usually Post inflammatory.

.....

arlond chiari malformation:

- ❖ congenital anomaly of brain in which cerebellar tonsils are elongated , pushed down through opening of base of skull (foramen magendi)
- ❖ usually hydrocephaly+- meningomyelocele

dandy walker malformation:

- ❖ congenital anomaly due cystic dilatation of ventricles cause obstruction of foramen magendi
- ❖ usually hydrocephaly+- cerebellar ataxia

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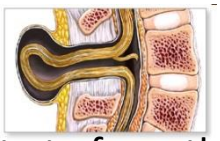
Neural tube defects



- ❖ Result from failure of normal fusion of the neural plate to form the neural tube during the first 28 days following conception.
- ❖ Due ↓ folic acid
- ❖ Detected: U/S and measurement of maternal serum and amniotic alpha-fetoprotein.
- ❖ Women who:

>may become pregnant are advised to get 400 MICg of folic acid daily.
>are pregnant → 1.0 mg
>have previously given birth to a child with a neural tube defect → get 4.0 mg

.....

ANENCEPHALY

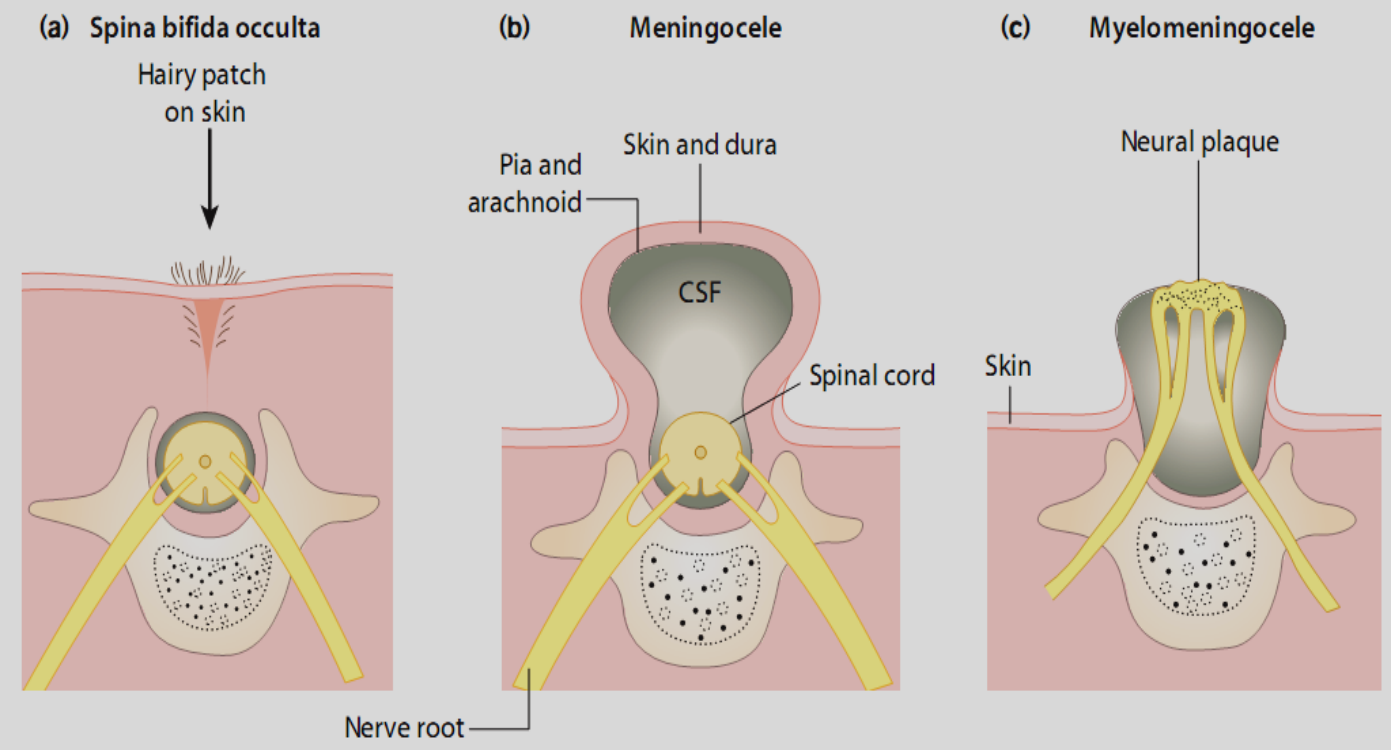
- ❖ Failure of development of most of the cranium and brain.
  Affected infants are still born or die shortly After birth.
- ❖ It is detected on antenatal ultrasound screening and termination of pregnancy is usually performed.

ENCEPHALOCELE:

- ❖ Extrusion of brain and meninges through a midline skull defect, which can be corrected surgically.
- ❖ There are often underlying associated cerebral malformations.

SPINAL BIFDA OCCULTA:

- ❖ This failure of fusion of the vertebral arch
- ❖ Is often an incidental finding on X-ray
- ❖ May be associated with overlying skin lesion such as a tuft of hair, lipoma, birth mark or small dermal sinus, usually in the lumbar region.
- ❖ There may be underlying tethering of the cord (diastematomyelia)→ may cause neurological deficits of bladder function,LL
- ❖ Can be diagnosed with MRI of spinal cord
- ❖ Neurosurgical relief of tethering is usually indicated.



SPINAL BIFDA CYSTA:

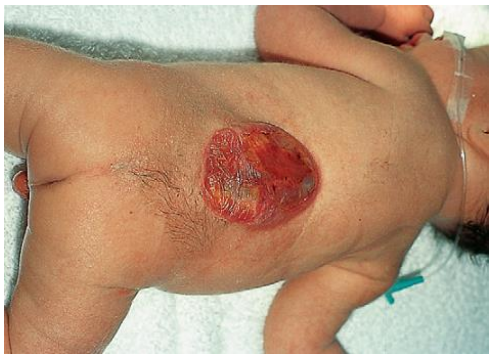
MENINGOCELE AND MYELOMENINGOCELE

Meningoceles→usually have good prognosis following surgical repair.

Myelomeningoceles may be associatedwith:

- Variable paralysis of the legs
- Muscle imbalance, which may cause dislocation of the hip and talipes
- Sensory loss

- Bladder denervation (neuropathic bladder)
- Bowel denervation (neuropathic bowel)
- Scoliosis
- Hydrocephalus from the Chiari malformation



Management:

- The back lesion is usually closed soon after birth.
- Paralysis and muscle imbalance –> physiotherapy- Walking aids-wheelchair
- Sensory loss→skin careis→avoid skin damage and ulcers.
- Neuropathic→ catheter be required

……

Small head (Microcephaly)

DEFINITION:

❖ OFC below 3th centile according to age and sex or 2 standards Deviation below mean

CAUSES:

	True microcephaly due to small sized brain	**Craniosynostosis**
Criteria	1. Skull sutures & fontanelles : normal. 2. No increase ICP. 3. Skull X ray show small vault. 4. CT scan show brain atrophy.	1- Palpable ridge is felt at the affected suture. 2- lf multiple sutures are affected→ brain atrophy. 3. Increase ICP→hydrocephalies beaten sliver appearance in skull X ray. 3- Skull examination →abnormal skull shape
causes	***A. Genetic:*** 1.Familial : AR 2.Chromosomal trisomy:21,18,13 ***B. Non genetic:*** • Prenatal: 1. TORCH infection.	due to early fusion of sutures

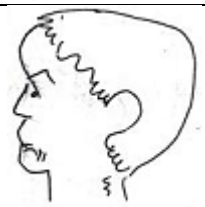
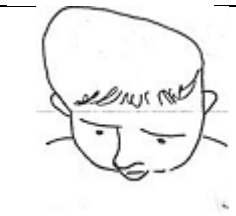
	2. Fetal irradiation. 3. Maternal diabetes or PKU 4.Maternal drugs Ex: phenytoin, alcohol. • Natal: HIE • Post natal: Early meningitis& Encephalitis	

TREATMENT:

1. Surgical separation of skull sutures is indicated in:

- Cases with hydrocephalus.
- Cases with progressively increase intra cranial tension.
- Cosmotic reasons.

ABNORMAL SKULL SHAPES:

name	shape	definition	Suture fused
scaphocephaly		Elongated naroow skull	Saggital suture
brachycephaly		Short Broad skull	Both coronal sutures
plagiocephaly		Unilateral forehead flattening	Single coronal or lambdoid
oxycephaly		High pointed head	Coronal Sagittal lambdoid

Cerebral palsy

DEFINITION:

2. Non progressive disorder of movement and posture involve immature brain of unknown etiology

ASSOCIATED WITH:

1. **MR** (2/3 of pts)
2. **Epilepsy** (1/3 of pts)
3. Impaired **hearing**
4. Impaired **vision**
5. Supranuclear bulber palsy:
   - A.Feeding disorder=**poor sucking,swallowing**
   - B.**Squint** (30%)
   - C.**Speech disorder**
   - D.**Persistence primitive reflex**

CAUSES:

Antenatal	Natal	Post natal
(8o%)	(10%)	(10%)
1. TORCHS 2. Cerebral dysgenesis 3. Fetal irradiation 4. Placental dysfunction 5. Maternal infection during pregnancy	1.HIE 2.Birth truma	1. IV hge 2. Meningitis ,encephalitis 3. Metabolic=PKU 4. Hypoglycemia 5. Hydrocephalus 6. Hyperbilirubinaemia

CLINICAL PICTURES:

1.Abnormal limb and/or trunk posture and tone
2.delayed motor milestones
3.may be accompanied by slowing of head growth
4.Feeding difficulties with oromotor incoordination,
5.slow feeding, gagging and vomiting
6.Abnormal gait once walking is achieved
7.Asymmetric hand function before 12 months of age

CLINICAL TYPES:

1. Spastic CP:

1. Common type 90%
2. Pyramidal tract lesion signs:
   - Hypertonia-Hyper reflexia
   - + Babinski and clonus

hemi	Arm and leg (arm>leg)
quadri	All limbs (arm>leg)
Di	All limbs (leg>arm)
mono	One limb
para	2 arms or 2 legs

Hemiplegia:

❖ Unilateral involvement of the arm and leg
❖ The arm is usually affected more than the leg, with the face spared.
❖ Affected children often present at 4–12 months of age with:

1. Fisting of the affected hand, a flexed arm, a pronated forearm, asymmetric reaching or hand function.
2. a tiptoe walk (toe–heel gait)
3. Affected limbs may initially be flaccid and hypotonic, but increased tone soon emerges as the predominant sign.
4. The past medical history may be normal, with an unremarkable birth history →with no evidence of hypoxic-ischaemic encephalopathy.

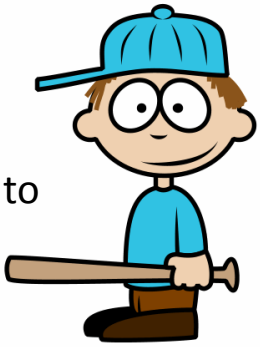
❖ In some, the condition its caused by neonatal stroke. Larger brain lesions (strokes)→may cause a hemianopia (loss of half of visual field) of the same side as the affected limbs

Quadriplegia:

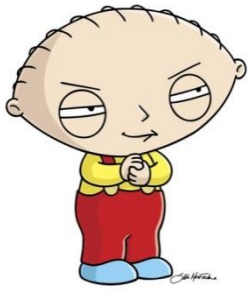
❖ all four limbs are affected
❖ sever form
❖ The trunk is involved with a tendency to opisothonus (extensor posturing), poor head control and low central tone (Fig. 4.5). This more
❖ is often associated with seizures, microcephaly and moderate or severe MR
❖ There may have been a history of perinatal HIE

Diplegia:

❖ all four limbs, but the legs are affected more than arms so that hand function may appear to be relatively normal.
❖ Motor difficulties in the arms are most apparent
❖ with functional use of the hands-Walking is abnormal.
❖ Diplegia is one of the patterns associated with preterm birth due to
❖ periventricular brain damage



2.Dyskinetic cerebral palsy(6%)

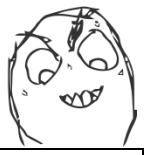


- ❖ Dyskinesia refers to movements which are involuntary,uncontrolled,
- ❖ more evident with active movement or stress.
- ❖ May be described as:
- • Chorea – irregular, sudden and brief non-repetitive movements
- • Athetosis – slow writhing movements occurring more distally
- • Dystonia – simultaneous contraction of agonist
- ❖ Intellect may be relatively unimpaired.
- ❖ Affected children often present with floppiness, poor trunk control and delayed motor development in infancy.
- ❖ Abnormal movements may only appear towards the end of the first year of life.
- ❖ Its due to damage in the basal ganglia or (extrapyramidal).
- ❖ In the past the commonest cause was hyperbilirubinaemia (kernicterus) due but it is now hypoxicischaemic encephalopathy at term.

3.Ataxic (hypotonic) cerebral palsy

- ❖ Most are genetically determined.
- ❖ Relatively symmetrical.
- ❖ There is early trunk and limb hypotonia, poor balance and delayed motor development.Incoordinate movements, intention tremor andan ataxic gait may be evident later.

4.atonic:



Floopy infant with herperreflexia

Investigation:	**Mangment:**
1.CT , MRI: •Detect degree of brain atrophy •May detect cause 2.Torchs screen and metabolic screen 3.EEG,audiometry,fundus examination	1. psychological support 2. Care feeding and defecation 3. Physiotherapy 4. Antispastic drugs: dantrolene ,baclofen ,botox 5. Assist vision and hearing 6. Assist walking =standing frames-wheel chair 7. Treat epilepsy 8. Rehabilitation according to MR degree

Seizures

DEFINITION:

- Clinical event in which there is sudden disturbance of neurological function caused by an Abnormal or excessive neuronal discharge

CONVULSIONS:

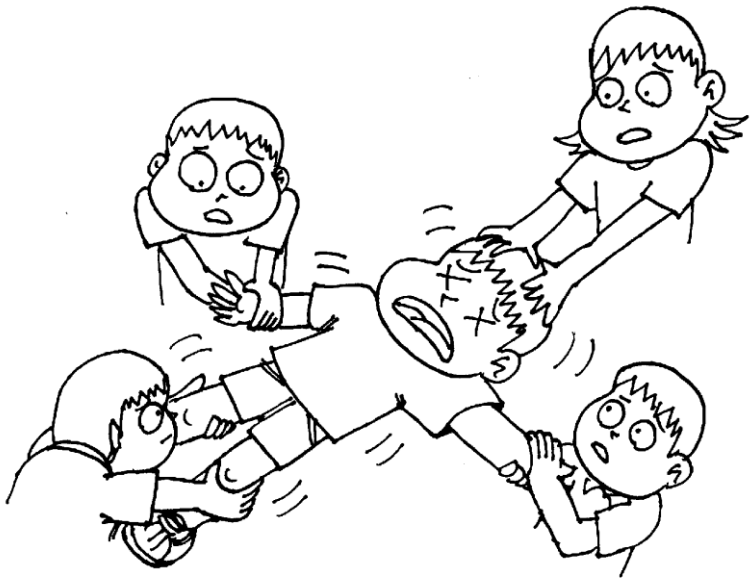
- Excessive abnormal muscle contractions , usually bilateral , that may be sustained or interrupted (motor seizures) .

CAUSES:

Acute convulsions:

1- Febrile convulsions.
2- First epileptic fit.
3- CNS causes:
- Infection→ meningitis, encephalitis, brain abscess.
- Irritation→ brain edema
- Tumors of the brain
- Toxic → tetanus or drug (EX: aminophylline)
- Hemorrhage→ trauma, hemorrhagic blood diseases.
- Hypoxia → HIE
- Hypertensive, uremic, or hepatic encephalopathy.

4- Metabolic causes:
- Hypo (glycemia, Ca, Mg)
- Hypo or hypernatremia
- Pyridoxine (B6) deficiency



Recurrent convulsions:

1- Epilepsy
2- Tetany
3- Degenerative brain diseases
4- Chronic metabolic causes:
- Hepatic encephalopathy
- Uremic encephalopathy.

Febrile Convulsion

DEFINITION:

- Seizures associated with fever in absence of another cause and not due to intra cranial infection.

INCIDENCE:

- ❖ 4% of children.
- ❖ Recurrent in 30-50% of cases.
- ❖ **+** F/H in 20% of cases (genetic base do exist).

CRITERIA FOR DIAGNOSIS:

1. Age 6m→6yrs (***not occur in neonate***).
2. Temperature → fever usually ↑39C°.
   (fit`s occur within 8-12hrs from onset of fever "early").
3. No evidence of CNS infection (meningitis, or abscess).
4. Evidence of extra cranial infection → (tonsillitis, otitis media, roseola).

You must ask about types in history:

TYPES:

Typical = simple	Atypical=complex
Generalized tonic clonic	Focal
Last ↓15min	Last ↑15 min
no recurrence in same illness	Recurrence occur in same illness
Commonest form	uncommon
❖ Does not affect intellectual ❖ Performance or risk of developing epilepsy ❖ There is 1–2%chance of developing epilepsy ,similar to the risk for all children	❖ increased risk of 4–12% of subsequent epilepsy

INVESTIGATIONS:

- **L.P.** → to differentiate from meningitis.(for complex type usually)
- **CT-MRI** → to differentiate from space occupying lesion
- **Blood sugar-Ca-Mg-Na**

- **EEG**→ is not indicated as it does not serve as a guide for treatment nor does it predict seizure recurrence.

Source: Illustrated.Textbook.of.Paediatrics.4th paper 472

D/D of fever+convulsion:

1. Febrile convulsion
2. Meningitis.
3. Viral meningoencephalitis.
4. Brain abscess.
5. Epileptic fit (precipitated by fever)

TREATMENT:

✓ Treatment the fever → paracetamol, or cold bath, or tepid sponges.
✓ Treatment the convulsion → rectal diazepam (if fits last more than 5 min.).
✓ Treatment the underlying cause → antibiotics.

- To reduce risk of recurrence at the onset of febrile illness give oral diazepam 0.3mg/kg/8h for 2-3days.
- Previous method is old and not used as they do not reduce the recurrence rate of seizures or the risk of epilepsy.

Source: Illustrated.Textbook.of.Paediatrics.4th paper 472

High risk group (to develop epilepsy):

1. Complex form
2. +F/H of epilepsy.
3. Pre-existing neurologic abnormality.
4. If developed before 9m.

Meningitis

DEFINITION:

- Inflammation of the membranes covering the brain & spinal cord.

TYPES:

1. Bacterial
2. Aspetic →viral(most common cause), fungal
3. Tuberculous (TB)

Bacterial (septic-pyogenic-purulent) meningitis

CAUSES:

Bellow 2 mts:	**Above 2 mts:**
1. streptococcus 2. staph aureus 3. G –ve (E coli) 4. Listeria monocytogenes	1. h. influenza (B) 2. strepto pneumonia 3. nesseria meningitis

Staph epidermises affect any age and can be due shunt

NOTE:

- Peak of H. influenza infection between 6-12 month --> incidence declined by vaccination
- Below 3 mts →low IgM is one of the commonest causes

TRANSMISSION:

1. Droplet infection mostly
2. Blood borne away in neonatal sepsis
3. Direct: OM-orbital cellulitis

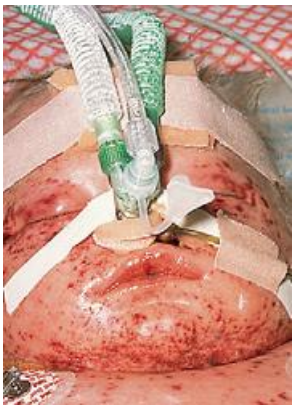
RISK FACTORS:

1. Male-black
2. Low immune as in AIDS-steroid use-cytotoxic drugs-chronic disease
3. Over crowded area
4. Meningomyocele-meningocele
5. Skull truma
6. Csf shunt
7. Anemia-spleen dysfunction

CLINICAL PICTURE:

1. Non specific

1. High fever (may be hypothermia in neonates).
2. Poor feeding
3. Rose spots (maculopapular rash)may appear on the trunk & extremities → meningococcal septicemia.



2. Features of increased ICP

Before fontanel closure:

1. Tense, bulging anterior fontanel
2. Irritable and poor feeding

After closure of fontanels:

1. Severe bursting headache (irritability)
2. Blurred of vision
3. Projectile vomiting (in the morning, not preceded by nausea)
4. Cushing response (hypertension & bradycardia)

3. Features of meningeal Irritation: (less sensitive in infants)

Neck rigidity (stiffness) → limited neck flexion

Opisthotonus→ arched back

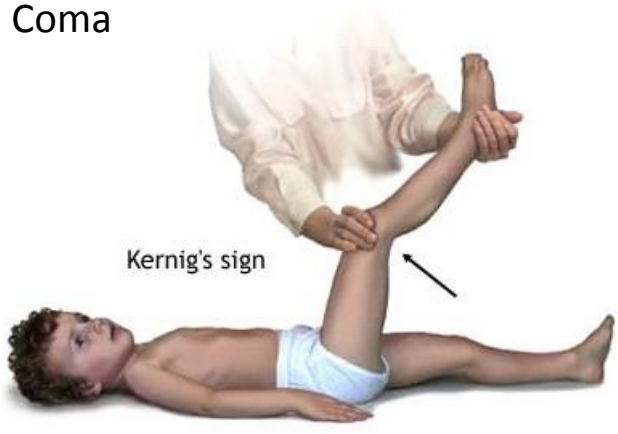
Kernig's sign → inability to extend the leg after the thigh is flexed to a right angle with the axis of the trunk.

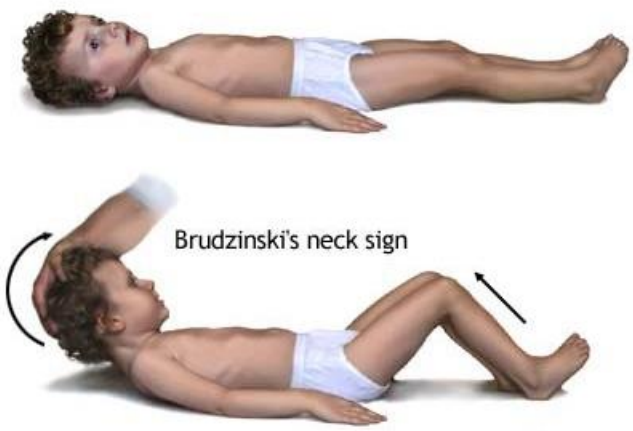
Brudzinski sign → Passive flexion of one hip → flexion of the other hip and knee

→ Passive flexion of the neck → flexion of the hip & knee.

4.Neurologlc signs :

1. Stupor & drowsiness.
2. Convulsions→ usually generalized
3. Coma





PATH PHYSIOLOGY:

Meningitis

Inflammation ↓	thrombosis of small blood vessels	damage of cerebral cortex ↓
Infiltration of inflammatory cells ↓		Obstruction of CSF pathway ↓
Pus ↓		CSF accumulation ↓
Thickening around brain and spinal cord		hydrocephaly

DIAGNOSIS:

- **In history:**

Ask about complain and analyse it (fever, convulsion).
Convulsion→Try to know if simple or complex and if there is a characteristic feature of febrile convulsion present. During analysis (preictal→ictal→post ictal)
Ask about recurrence and history of trauma and epilepsy in family In systemic review →try to find septic focus in case of febrile convulsion

- **In examination:**

Manifestations of ↑ICP .
Manifestations of meningeal irritation.

- **Investigations:**

1- CBC→leucocytosis

2- CRP-ESR→+

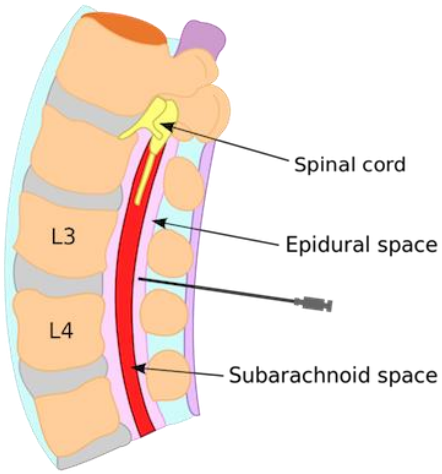
3- CSF (LP):

iF pt

↓2yrs	↑2yrs
Mandatory L.P. Examine the pt to find if there is any septic focus.	If meningeal irritation Present (+) → Do L.P. not present (-) → treat pt as febrile convulsion

- How to do **L.P**.? (oral very important).
- If pt sever ill, shocked → do complete septic screen :
- For complication → CT/MRI for head.

1. Blood C/S.
2. Urine C/S.
3. Chest X-ray.
4. L.P.
5. Throat swab for C/S.



Result of L.P.

	normal	bacterial	viral	T.B.
Colour	Clear	Turbed	Clear	Clear
Cells pus (WBC)	0-5/mm³	200-5000↑↑↑	100-700 normal or ↑	100-500 ↑↑
Type of cells	Lympho.	PMN	Lympho.	Lympho.
Protein	15-35mg/dl	↑↑↑	↑	Normal or slight ↑
Sugar	⅔ or 60% of serum sugar	Low	Normal or low	Low

TREATMENT:

1- *Empirical antibiotics (anti-meningitic dose):*

If pt

↓2m	↑2m
(1st line): Ampicillin + Gentamicin ↓ ↓ Strepto E.coli group β ↑ ↑ lesteria (best for it Ampicillin) If Staph. → Genta.+Cloxacillin ***(2nd line):*** Ampicillin + 3rd generation Ceftriaxone Cefataxine	3rd generation cephalosporin for: - H. influenza. - N. meningitis. - Strepto. pneumonia.

- if pt → infant 2-3 week.
  →child 10-14 day.
- by IV rout.
- if there is H.influanza give **Dexamethazone** to prevent deafness→ Beyond the neonatal period,dexamethasone administered with the antibiotics reduces the risk of long-term complications such as deafness .

2- *Supportive treatment:*

1. Antipyretic→paracetamol
2. Care of feeding → IV fluid.
3. Anticonvulsion→immediate relief diazepam then phenobarbitone
4. ↓ ICP → manitol—furosemide-hyperventilation
5. Steroids → incase of: h.influenza-septic shock-adrenal faliure

3- Treat complications.

COMPLICATION:

early	Late
1- Convulsion. 2- ADH → oedema. 3- Subdural haemorrhage, or effusion. 4- Shock. 5- DIC. 6- Brain abscess.	1- Epilepsy. 2- Mental retardation. 3- Deafness (30%)→ **H.influenza due to 8th cranial damage.** 4- Hydrocephaly. 5- Learning disabilities. 6- Optic neuritis.

PREVENTION:

- Isolation of case→side room
- Rifampicin to all family members especially in H. influenza because it spread by Air droplets.
- Any case at (admission, discharge, or follow up) → measure O.F.C.
- Follow up at least 2 years:

1. Developmental assessment (hearing, vision).
2. Neurological examination.

- Vaccine:

1.H. influenza β vaccine (3doses)-meningococci-pneumococci
2.Meningococcal polysaccharide vaccine (A, C).

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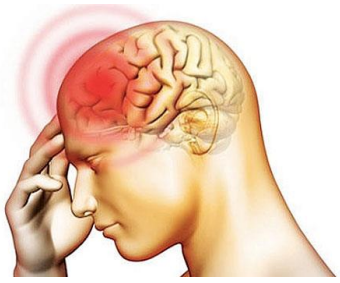
viral = aseptic = atypical (it`s the most common cause)

- Meningitis with no micro organism detected in CSF by gram stain or bacterial culture.

A.Viral→ aseptic = atypical (it`s the most common cause)



1.HsV.
2. enteroviruses→echo and coxachie (most common cause)
3.EBV.
4.Mumps .

B.Protozoal → toxoplasma-malaria.

C.Non infectious→ cns leukemia-Intrathecal injection-Post vaccination

Treatment :Supportive and antiviral

Encephalitis

DEFINITION:

- inflammation of the Brain.

CAUSES:

1. **Enterovirus**→most common cause
2. **Herpes simplex** virus (HSV 1.6.7)
3. **Arboviruses**
4. **Epstein-Barr virus** (EBV),
5. **HIV**
6. mononucleosis' virus associated with childhood illness (mumps/measles/rubella/varicella).
7. CMV → cytomegalovirus.
8. Rabies.
9. Bacterial as: Mycoplasma, Borrelia burgdorferi (Lyme disease), Bartonella henselae (cat scratch disease), rickettsial infections (e.g. Rocky Mountain spotted fever)

CLINICAL PICTURES:

1.Early manifestation (viremia):
(fever, headache, vomiting).
2.Neurological manifestations develop suddenly → instable level of conscious which may range from →irritability, confusion → to deep coma.
3.↑ICP manifestations.
4.Specific aetiology manifestations.

DIAGNOSIS:

1.**Lumbar puncture** (maybe normal)

- High cells →mainly lymphocyte pleocytosis 10-500 cell/m^3.
- High protein, normal glucose.

2.**EEG**→definitive test→slow wave activity
3. **C.T-scan or MRI**: mandatory

- May detect focal or generalized abnormality in pt with encephalitis.

(temporal lobe focus on CT or EEG→herpes simplex)

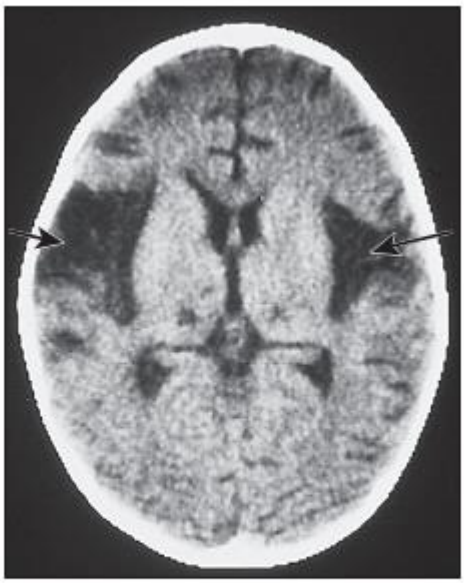
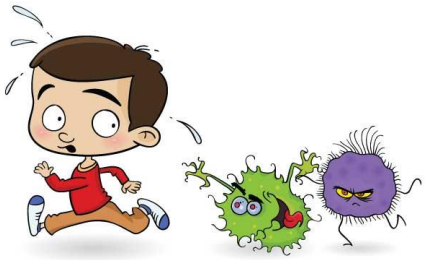


Figure 14.5 Herpes simplex encephalitis. The CT scan shows gross atrophy from loss of neural tissue in the temporoparietal regions (arrows).

TREATMENT:

Supportive treat:



1. Pt. direct admission in ICU.
2. Decrease ICP or
3. ↓risk of cerebral oedema by:

-Head elevated in 30° → in neutral position.

-Reduction of fluid intake to 70% of fluid requirement.

-Drugs like →lasix, mannitol, Dexamethazone (steroids) →↓ICP.

4.Mechanical ventilation in severe cases.

Antimicrobial therapy for herpes simplex, encephalitis → Acyclovir for 10 day.

*D/D of fluid intake restriction:

1- D.K.A. (slow correction) diabetic ketoacidosis.
2- CHD. Congenital heart disease.
3- Acute renal failure.
4- Birth asphyxia.
5- Encephalitis.
6- Meningitis.
7- Hypertonic dehydration (slow reduction).

Epilepsy

DEFINITION:

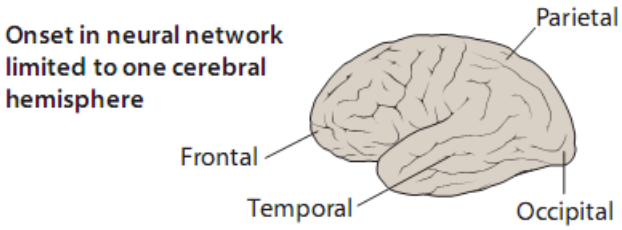
- chronic neurological disorder characterised by recurrent unprovoked seizures that occur at interval greater than 24 hrs

CAUSES-TYPES:

type	*Idiopathic (primary)*	*organic (secondary)*
%	80% of cases	20% of cases
causes	Genetic basis exist for many epileptic syndromes	1. Congenital cerebral malformation 2. Degenerative brain diseases. 3. post-traumatic. 4. Post-hemorrhagic. 5. Post-infection. 6. Post-toxic. 7. Post-anoxic.

CLASSIFICATION:

1. Focal (partial) seizures
2. Generalized seizures
3. Partial with 2ry generalized

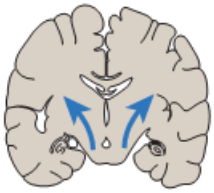


FOCAL (PARTIAL) SEIZURES:

- Only one part (side) of the body is involved=one hemisphere

1.Simple partial seizures	*2.Complex partial seizures*
Benign rolandic epilepsy or benign childhood epilepsy	
No aura	Preceded by aura (Ex: headache)
May be motor, sensory , autonomic	Only motor fits
No automatism Consist of twitching or jerking of one side of face-arm-leg Excellent prognosis	Automatism may occur → automatic behaviors as chewing, suckling, lip smacking or aggressive actions as rubbing ,pulling of clothing
Consciousness is intact	Consciousness is impaired.
Common in boys, at night-Age:5-10 yrs	
EEG:centrotemporal spikes	EEG:Frontotemporal spikes

GENERALIZED SEIZURES:



- The whole body is affected(both hemisphere)

1- Absence seizures (petit mal):

Incidence:

- ❖ More in girls
- ❖ 5% of epilepsy
- ❖ common above 5 years(5-12 yrs)

Atypical type:
More than 30 sec
Involuntary movement(myoclonic)
EEG:rapid irregular waves
Prognosis:not good

Description:

- ❖ Sudden cessation of all motor activities or speech with a blank facial expression and flickering of eye lids without falling down or abnormal movments

Precipitated by hyperventilation for 3-4 min g or photic stimulation.

- ❖ Last < 30 seconds→ after seizure patient resume preseizure activity.
- ❖ Impairment of consciousness is the essential symptom, and may be the only clinical symptom
- ❖ Frequently recurrent with **No aura**
- ❖ **EEG :** typical 3/second spike and generalized wave discharge.
- ❖ **Prognosis:** good→40% develop generalized tonic clonic

2- Generalized tonic clonic seizures (Grand mal):

- ❖ The commonest form; pass in 3 phases.

Aura=pre ictal	ictal	Post ictal
Warning sign before attack: **Motor**→spasm **Sensory**→paraesthesia **Autonomic**→abdominal pain	loss of consciousness • **Tonic phase:** Tonic contraction of whole body→rigid posture ,apnea,cyanosis,rolling of eyes,drolling of saliva • **Clonic phase:** Rhythmic contraction ,relaxation of all muscle group→tongue biting, loss of sphincter control	Semiconscious for 30 min →2hrs Headache sleep

3- Myoclonic epilepsies:

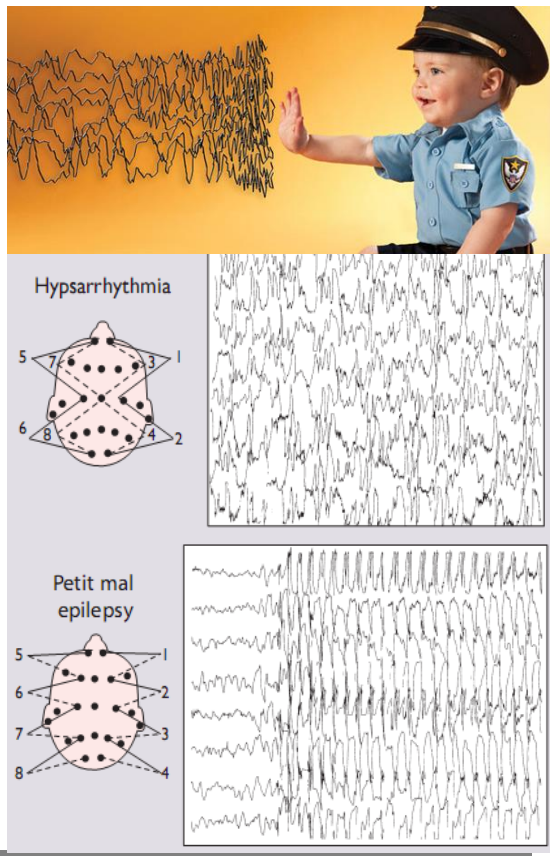
- Sudden shock like repetitive contractions of group of muscles → with loss of body tone.
- Intact consciousness.
- EEG →4-6 spike/sec. irregular polyspike waves.

4- Infantile spasms:

- Also called west syndrome- salaam spasms
- Starts in the 1th year of life→ peaking between 4- 8 months
- Brief symmetric tonic contractions of the neck ,extremities & trunk
- Which may be flexor, extensor or mixed
- Repetitive→ usually in the morning → on waking
- A cry may precede or follow the spasm→so may be confused with colic
- May be associated with developmental delay (West syndrome)
- EEG → commonly show **hypsarrythmias** (irregular, high amplitude waves)
- Cause→ 2\3 have underlying neurological cause (2ry cause is the common cause)→ Main tuberous sclerosis-IEOM-asphyxia
- Treatment→ with vigabatrin or ACTH or corticosteroids→good response in 30–40% but unwanted effects are common→Most will subsequently lose skills
  And develop learning disability or epilepsy

5- Atonic seizures:

- Sudden loss of body tone.



INVESTIGATION:

1- **EEG** →not diagnostic (normal between attacks)
2- **Metabolic screen**→ Serum Na, Ca, Mg, glucose
3- **CSF** →in suspected CNS infections.
4- **CT brain in**:
- Focal lesions
- Increased intra cranial pressure
- Resistance to treatment
5- **Serum anticonvulsant level** for:
- At the onset of anti convulsant therapy to confirm therapeutic range
- Polytherapy- Drug toxicity
- Status epilepticus

D-D:

1. Syncopal attacks: -

- Fainting with loss of consciousness due to brief brain ischemia.
- Due to vagal stimulation or arrythmia

2- Breath holding attacks: page 45

3- Hysterical fits

TREATMENT OF EPILEPSY:

a. Advise the parent → to watch the child during swimming, passing traffic, ...
→ never to stop the antiepleptic drug suddenly

b. Anti-epileptic drugs :

- Only one drug is used with small dose → if no response gradually increases the dose

- in resistant cases → 2nd drug can be used alone or in combination.
- Duration of treatment is at least 2 years after last attack

D.O.C

Simple partial→ Carbamazepine= Tegretol
Complex partial→ Carbamazepine= Tegretol
Absent seizures→ Ethosuximide
Atypical absent→Na valproate
Infantile spasm→ACTH-steroid- Vigabatrin
Generalized tonic conic→Na valproate

Side effect:

Valproate	Weight gain, hair loss Rare idiosyncratic liver failure
Carbamazepine	Rash, neutropenia, hyponatraemia, ataxia Liver enzyme induction, can interfere with other medic
Vigabatrin	Restriction of visual felds, which has limited its use Sedation-Lamotrigine Rash
Ethosuximide	Nausea and vomiting
clonazepam,diazepam	Sedation, tolerance to effect, increased secretions
All the above may cause drowsiness and occasional skin rashes.	

Role of community:

1. Regular follow up→LFT-RFT
2. Don't stop antiepileptic drugs suddenly→withdrawal→status epilepticus
3. Don't jump from Dr to another
4. Avoid risk→driving-swimming-sharp instruments-long time on Tv-playstation
5. Psychological and reassurance.

70% of pts are free of seizures at 16 years old

.....

Status epilepticus

DEFINITION:

☒ seizure lasting 30 minutes or longer or when successive seizures occur so frequently that the patient does not recover consciousness between them

CAUSES:

1. Prolonged febrile seizures (common cause)
2. Sudden withdrawal of antiepileptic drugs
3. CNS anomaly Ex: tumers-encephalitis
4. Metabolic Ex: hypoglycemia
5. IEOM

MANGMENT:

1. Stabilize vital functions

Call for help

A=cleaning air way →put baby in left lateral position-suction

B=give O2 by mask + monitor o2 saturation

C=insert cannula and support circulation by IVF if case need and take blood for Investigation as above

2. Correct transient metabolic disturbance

1- Low glucose→dextrose water 10% IV state

2- Low CA→Ca gluconate slow IV

3- Low Mg→ mg sulphate 50% IM

3. Anticonvulsant agent if seizure prolonged ↑ 5min or repeated

Note:
Some protocols
(as in our hospitals) we advise to stop convulsion immediately without waiting by diazepam or buccal midazolam

Phenobarbitone(in infant)
Loading dose= 20mg/kg slow IV
Maintenance dose=3-8 mg/kg IV

If no response add

Phenytoin
Loading and Maintenance dose as Phenobarbitone

If no response add

Midazolam or clonazepam

If no response add

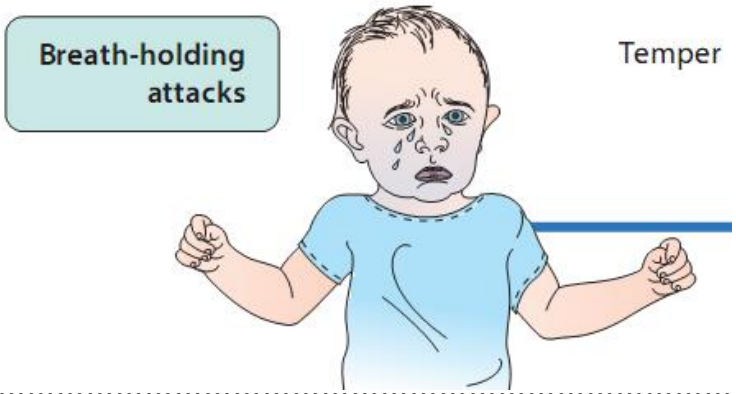
Pyridoxine 50 mg IV

GA+MV

After attack treat under line cause and gradual withdrawal of anticonvulsant

Breath-holding attacks

☒ Crises→crying→hold breath→cyanosis→ Fainting or loss of alertness (unconsciousness)→rapidly recover
☒ Onset=3ths→6 yrs (common in toddler
☒ Two types=blue-pale types
☒ Benign condition→ Drug therapy is unhelpful. Attacks resolve spontaneously,but behaviour modification therapy, with distraction, may help



SIDS
sudden infant death syndrome

☒ Unexplained death by history or through post mortem examination in infancy
☒ Death mainly at mid night up to 9 am
☒ More in winter
☒ Mainly 2 mtd → 3mts

RISK FACTORS:

1- Preterm
2- Lack of perinatal care
3- Placing an infant to sleep while lying on the stomach or the side↑the risk
4- Maternal smoke during pregnancy
5- LSES
6- Parental smoke
7- Elevated or reduced room temperature
8- idiopathic

The neurocutaneous syndromes

☒ The nervous system and the skin have a common ectodermal origin

NEUROFBROMATOSIS TYPE 1 (NF1)

☒ Commonest neurocutaneous syndromes
☒ Affects 1: 3000 live births.
☒ Autosomal dominant
☒ One-third have new mutations.
☒ To make the diagnosis two or more of these criteria need to be present:

1. Six or more café-au-lait spots >5 mm in size before puberty (>15 mm after puberty)
2. More than one neurofibroma,firm nodular overgrowth of any nerve
3. Axillary freckles
4. Optic glioma→visual impairment
5. One Lisch nodule,a hamartoma of the iris seen on slit-lamp examination
6. Bony lesion from sphenoid dysplasia,which can cause eye protrusion
7. First-degree relative with NF1.

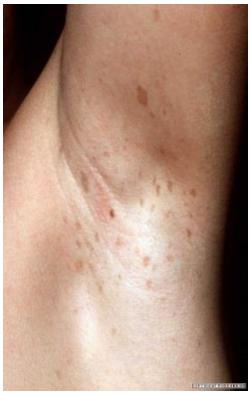
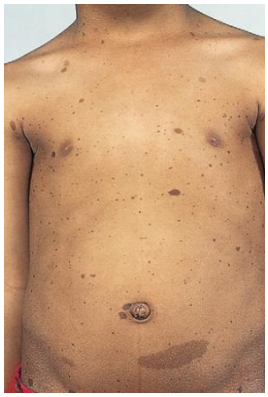
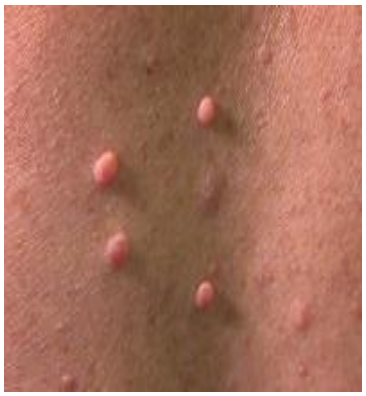
Associated problems:

1- Visual or auditory impairment→ compression of the 2th or 8th cranial nerve
2- optic glioma
3- pheochromocytoma
4- neuroblastoma
5- learning disability
6- epilepsy
7- macrocephaly
8- pulm HTN
9- renal artery stenosis +HTN
10- MEN syndrome

D-D of café-au-lait spots:

1- Neurofibromatosis
2- Ataxia telangiectasia
3- Tuberous sclerosis
4- MENs
5- Noonan syndrome
6- Russel silver syndrome

☞ *However, most people with the disorder carry no features other than the Cutaneous stigmata*

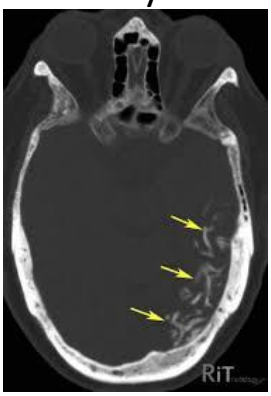
STURGE–WEBER SYNDROME

☒ sporadic disorder with a haemangiomatous facial lesion (a port-wine stain) in the distribution of the trigeminal nerve associated with a similar lesion intracranially.

☒ The ophthalmic division of the trigeminal nerve is always involved

☒ xray→show 'rail-road track' due Calcification

☒ MRI →best choice



Associated with:

1- Epilepsy
2- Learning disability
3- Hemiplegia.
4- There is high risk of glaucoma

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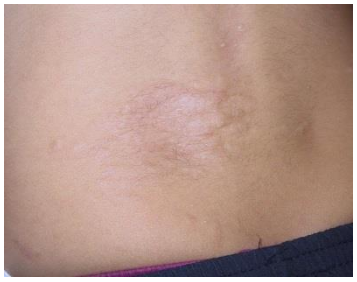
TUBEROUS SCLEROSIS:



☒ Autosomal dominant

☒ Up to 70% with new mutations.

☒ 1:9000 live births.

The cutaneous features consist of:

1- Depigmented 'ash leaf 'shaped patches which fluoresce under ultraviolet light (Wood's light)

2- Roughened patches of skin (shagreen patches) → over the lumbar spine

3- Adenoma sebaceum (angiofibromata) in a butterfy distribution over the bridge of the nose and cheeks,which are unusual before the age of 3 years

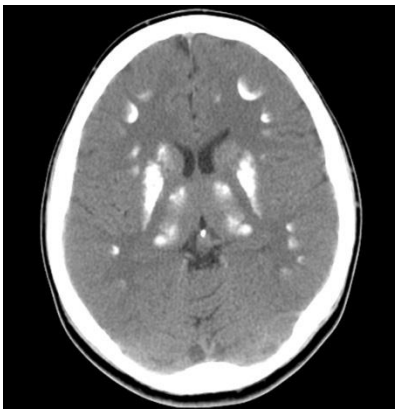
Neurological features are:

1- Infantile spasms and developmental delay
2- Epilepsy–often focal
3- Intellectual impairment.
4- autistic features
5- Fibromata beneath the nails (subungual fibromata)
6- Rhabdomyomata of the heart
7- Polycystickidneys.
8- Dense white areas on the retina (phakomata) from local degeneration

☒ CT-MRI→ detect the calcifed subependymal nodules and tubers from the second year of life.

D-D OF INTRACRANIAL CALCIFICATION:

A.Neurodermatosis (Tuberous sclerosis- Sturge–Weber syndrome)
B.Trauma
C.Inflammation:
1- Congenital toxoplasmosis
2- CMV
3- Rubella infection
4- Pyogenic abscess
5- HSV
6- Echinococcus



D.Tumor:Craniopharyngioma-Teratoma-Astrocytoma
E.Metabolic causes:Vit D exess-Hypo and hyper parathyroidism

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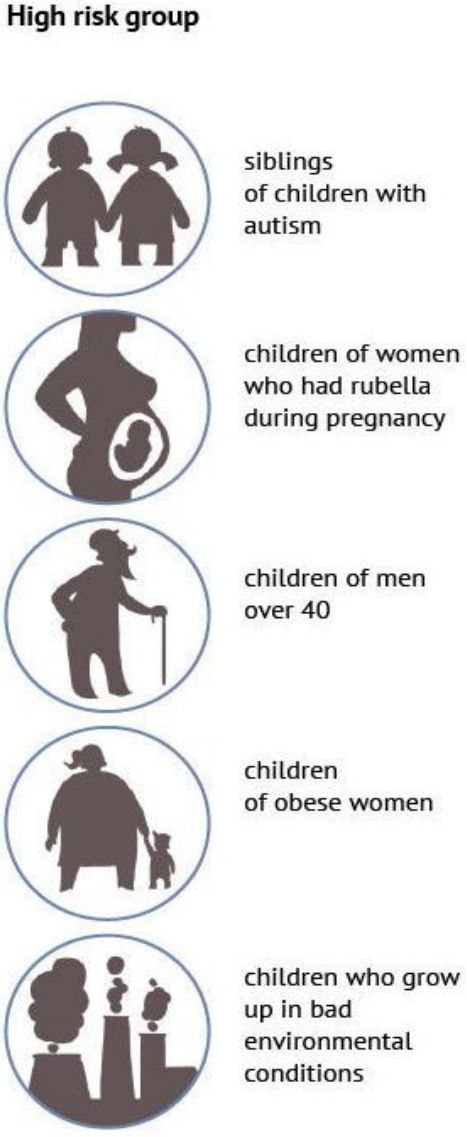
Autistic spectrum disorders

☒ Its disorder of neural development that is characterized by impaired social interaction and communication
☒ **3–6/1000** live births
☒ Common in **boys (4:1)**
☒ Increased in **industrial area**
☒ Has strong **genetic basis**
☒ it's not the result Of emotional trauma, Deviant parenting.

☒ Presentation is usually between 2 and 4 years of age when language and social skills normally Rapidly expand
☒ There is no evidence for a Suggested link with The MMR vaccine.
☒ **Asperger syndrome:** refers to a child with the social impairments of an autistic spectrum Disorder but at the milder end,and near Normal speech development.



☒ Autism is diagnosed by observation of behaviour,Including the use of formal standardised tests.

> ■ It is difficult to diagnose children under two years old, as very young children can develop at substantially different rates

1. Impaired social interaction:

- does not seek comfort, share pleasure form close friendships
- prefers own company, no interest or ability in interacting with peers (play or emotions)
- gaze avoidance
- lack of joint attention
- socially and emotionally inappropriate behavior
- does not appreciate that others have thoughts and feelings

tiptoes or flaps hands

3. Speech and language disorder:

- Delayed development, may be severe
- Limited use of gestures and facial expression
- Formal pedantic language, monotonous voice

3.ritualistic and repetitive behavior:

- On self and others, with violent temper tantrums if disrupted
- Unusual stereotypical movements such as hand fapping and tiptoe gait
- Peculiar interests and repetitive adherence

4.Co-morbidities:

- General learning and attention difficulties (about two thirds)
- Seizures (about one quarter, often not until adolescence).

avoids eye contact, doesn't gesticulate, doesn't change facial expression

repeats certain words and phrases or speaks about him or herself in second or third person

doesn't answer to his or her name, is frightened by certain sounds or has other "inexplicable" phobias

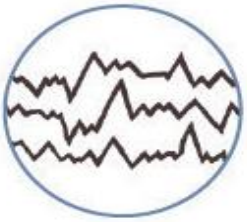
Management

☒ Usually managed by behavior modification such as applied behavioral analysis (ABA).

What to do if you suspect your child has autism:

See a neurologist to rule out any diseases related to brain development anomalies

See a children's psychiatrist



An EEG, an MRI, a hearing test and a Doppler echocardiogram may also help with diagnosis

A team of specialists should carry out the examination

- A psychologist
- A neuropathologist
- A children's psychiatrist
- A pediatrician
- A speech therapist
- Other experts in children's special needs

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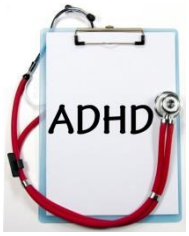
Attention deficit hyperactivity disorder (ADHD)



☒ the child is overactive in most situations and has impaired concentration with a short attention span or distractibility.

☒ 10-50:1000

☒ Boys**>** girls x3.

☒ There is a powerful genetic predisposition→dysfunction of brain neuron circuits that depend on dopamine as a neurotransmitter and which Control self-monitoring and self-regulation.

Presentation:

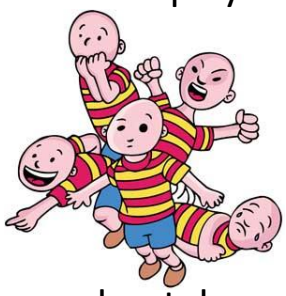
☒ Children are unable to sustain attention or persist with tasks.

☒ They cannot control their impulses

☒ they manifest dis-organized, poorly regulated and excessive activity

☒ have difficulty with taking turns; sharing

☒ Socially disinhibited; and butt into other people's conversations and play with poor relationships with other children.



☒ The children do poorly in school

☒ Variety of reasons→drives parents,teachers→punishment

Management:

A.Education and behaviour:

☒ in preschool children and school-aged children with mild to moderately Disorder→ active promotion of behavioral and educational progress by specific advice to parents and teachers→These involve having clear rules and expectations

B.medications:

☒ for children older than 6 years of age

☒ Ex:methylphenidate or dexamphetamine and atomoxetine

☒ reduce excessive motor activity and improve attention on task

☒ approach is not to put the child on medication until behavioral and educational progress is actively promoted

C.diet:

☒ Current evidence indicates that the sort of diet which aims blindly to reduce sugar,artificial additives or colourants has no effect.

☒ exclusion of some type of foods (which may show excitability or irritability)

☒ In general, food and drinks with caffeine are not advised.

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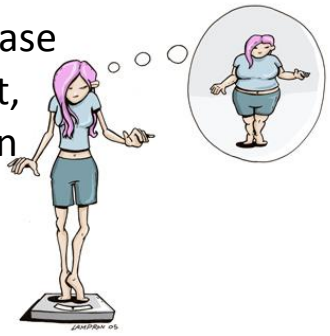
Eating Disorders

Essentials of Diagnosis:

Anorexia nervosa (AN)	Bulimia nervosa
body weight <85% of expected from height,	body weight usually normal
1. fear of weight gain, disturbed body image, denial, attempts to camouflage thinness, amenorrhea 2. lanugo, dry skin 3. hyponatremia 4. bradycardia 5. hypokalemia **6.** hypothermia	1.episodic binge eating with inappropriate compensatory behavior (emesis, laxatives, enemas, diuretics, diet pills,fasting, excessive exercise) 2.sense of being out of control during and after attacks 3.disturbed body image

Treatment:



- Early intervention and education may prevent full-blown disease
- Recognize the early signs of anorexia—sudden intense low fat, low carbohydrate diet, complaints of early satiety, failure to join family meals, intense concern over body image, unexplained weight loss, amenorrhea
- Unexplained disappearance of groceries may alert parents to bulimia
- Team approach to treatment is most successful with nutritional monitoring, education, family counseling, psychiatric evaluation and treatment, and medical subspecialists as needed
- Hospitalization may be needed.
- **Mortality in AN from suicide,electrolyte disturbance, or cardiac arrhythmia is up to 18% depending upon disease severity**

……

Central motor disorders

Corticospinal (pyramidal) tract disorders	Basal ganglia disorders	Cerebellar disorders
Cerebral dysgenesis, e.g. neuronal migration problem	Acquired brain injury: – Acute and profound hypoxia-ischaemia – Carbon monoxide poisoning – Post cardiopulmonary bypass chorea	Acute – medication and drugs, including alcohol and solvent abuse
Global hypoxia-ischaemia		Post-viral – particularly varicella infection
Arterial ischaemic stroke		Posterior fossa lesions or tumours, e.g. medulloblastoma
Cerebral tumour		Genetic and degenerative disorders, e.g. ataxic cerebral palsy, Friedreich ataxia and ataxia-telangiectasia
Acute disseminated encephalomyelitis		
Post-ictal paresis		
Hemiplegic migraine		
	Post-streptococcal chorea (rheumatic fever)	
	Mitochondrial cytopathies	
	Wilson disease	
	Huntington disease	

Cerebrovascular disease

Intracranial haemorrhage

Extradural haemorrhage	Subdural haematoma	Subarachnoid Hge
Follows direct head trauma, skull fracture	characteristic lesion in non-accidental injury caused by shaking or direct trauma ininfants or toddlers	The cause is often an aneurysm or arteriovenous malformation (AVM).
Tearing of middle meningeal artery as it passes through the foramen spinosum of the sphenoidbone	Tearing of the veins as they cross the Subdural space	
1. lucid interval conscious level deteriorates 2. seizures 3. increasing size of the 4. haematoma 5. focal neurological signs 6. with dilatation of the ipsilateral pupil 7. paresis of the 8. contralateral limbs and 9. anaemia 10. shock	Retinal haemorrhages Hypoxia Subdural haematomas	acute onset : head pain neck stiffness occasionally fever. Retinal haemorrhage is usually present. Seizures and coma may develop
Confrmed with CT scan		1.CT scan of the head 2.blood in the CSF
1.Correct hypovolaemia, 2.urgent evacuation of the haematoma And arrest of the bleeding		Treatment can be neurosurgical or with Interventional radiography

Stroke

☒ Occur in infants and children

☒ Causes include:

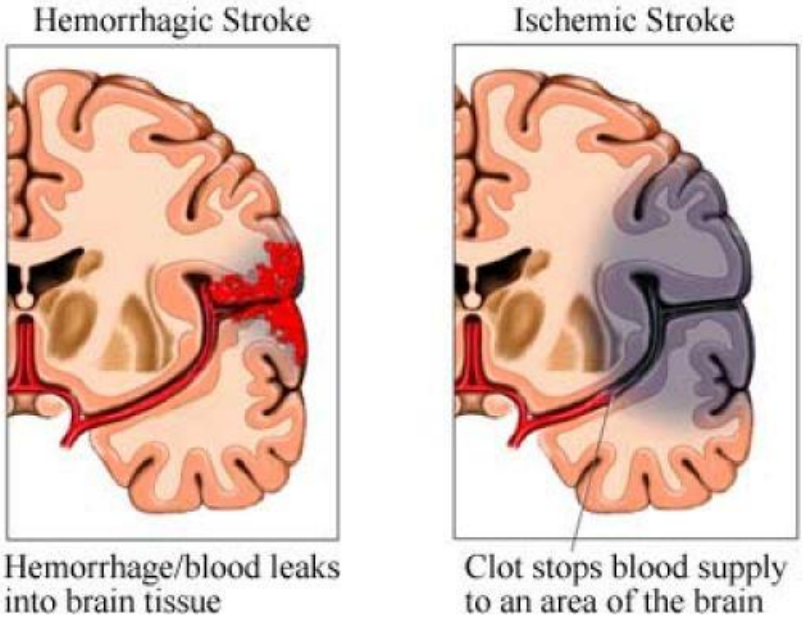
- **Cardiac:** CHD,Ex:FOT-endocarditis
- **Haematological:** sickle cell disease,deficiencies of anti-thrombotic factors,
- **Post-infective:**following varicella or other viral infection
- **Infammatory:** damage to vessels in autoimmune disease Ex: SLE
- **Metabolic/genetic:**homocystinuria,mitochondrial disorders Ex:**MELAS** (**m**yoclonic **e**pilepsy,**l**actic **a**cidosis and **s**troke)
  CADASIL (**c**erebral **a**utosomal **d**ominant **a**rteriopathy with **s**ubcortical **i**nfarcts And **l**eukoencephalopathy)→the most common form of hereditary stroke disorder
- **Vascular malformations:** moyamoya disease.

The clinical presentation:

1. Hemiparesis with or without speech disturbance.
2. Less common is compromise of the posterior circulation→due to compromise of the anterior circulation (internal carotid,anterior and middle cerebral arteries)
3. visual or cerebellar signs→due to compromise of the posterior circulation (vertebrobasilar arteries)

Investigations:

1. MRI
2. carotid Doppler
3. angiography
   to detect cause Ex:
4. echocardiography→embolism thrombophilia and vasculitis screen, and metabolic

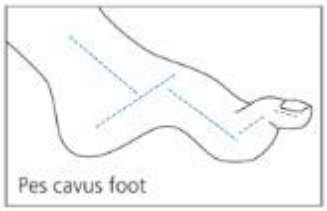


management:

1. Rehabilitation requires
2. The involvement of the remedial therapy team.
3. Aspirin prophylaxis is recommended
4. anti-thrombolytic agents.

Ataxia
Friedreich ataxia

☒ autosomal recessive.
☒ It presents with:
1. worsening ataxia
2.distalwasting in the legs
3. absent lower limb reflexes but extensor plantar responses because of pyramidal involvement
4.dysarthria,pes cavus
5.Impairment of joint position and vibration sense
6.optic atrophy.
7.cardiomyopathy→cardiorespiratory compromise→deathat40–50 years.



Ataxia telangiectasia

☒ autosomal recessive→ disorder of DNA repair
☒ It presents with:
1.delay in motor development in infancy
2.oculomotor problems→ incoordination and (oculomotor dyspraxia)
3.diffculty with balance and coordination becoming evident at school age.
4.Many children require a wheelchair for mobility
5. Telangiectasia develops in the conjunctiva,neck and shoulders fromabout 4 years of age.

<u>These children:</u>

- increased susceptibility to infection→principally from an IgA surface antibody defect
- Develop malignant disorders, principally acute lymphoblastic leukaemia (about 10%)
- raised serum alpha-fetoprotein
- increased WBC sensitivity to irradiation, Which can be used diagnostically, but The ATM gene test is now mostly used

Mental Retardation

DEFINITION: Handicapping disorder with age of onset below 18 years characterized subnormal I.Q. (< 70%).

I Q (intelligence quotient)= mental age x100
Chronological age

DIAGNOSTIC CRITERIA:

1- Subnormal intelligence quotient (less than or equal to 70%)
2- Limitations exist in two or more of the adaptive skills Ex:
communications, social skills, self care, safety, functional academics, work
3- Manifest before age of 18 yrs if after 18 years, it is called dementia

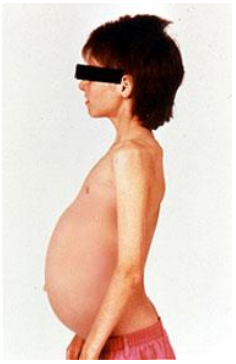
CAUSES:

Non-organic causes (physiological)
- In about 80-90% of cases.
- Usually mild
- No demonstrable brain abnormality.

organic (pathological group)
1. Chromosomal anomalies→ Trisomy 21,18,13, klinefelter syndrome
2. Genetic disorders → Fragile-x syndrome , prader willi syndrome
3. Cerebral palsy
4. Developmental brain abnormalities → hydrocephalus
5. Inborn errors of metabolism
6. Familial retardation (environmental , genetic)
7. Congenital infections
8. Congenital hypothyroidism

Neurodegenerative disorders

- These are disorders that cause a deterioration in motor and intellectual function. Abnormal neurological features develop, including:
  1. Seizures
  2. Spasticity
  3. abnormal head circumference (macro-or microcephaly),
  4. Involuntary movement disorders
  5. visual and hearing loss and Behaviour change.
- This disorders includes:



- Lysosomal storage disorders,e.g.lipid storage disorders and mucopolysaccharidoses
- Peroxisomal enzyme defects,Ex: X-linked adrenoleucodystrophy (VLCFAs)
- Heredodegenerative disorders,Ex: Huntington disease,which presents With progressive dystonia,dementia,seizures and corticospinal tract signs

Table 27.5 Lipid storage disorders

Disorder	Enzyme defect	Clinical features
Tay–Sachs disease	Hexosaminidase A	Autosomal recessive disorder Most common among Ashkenazi Jews Developmental regression in late infancy, exaggerated startle response to noise, visual inattention and social unresponsiveness Severe hypotonia, enlarging head Cherry red spot at the macula Death by 2–5 years Diagnosis – measurement of the specific enzyme activity Carrier detection of high-risk couples is practised Prenatal detection is possible
Gaucher disease	Beta-glucosidase	Occurs in 1 in 500 Ashkenazi Jews Chronic childhood form – splenomegaly, bone marrow suppression, bone involvement, normal IQ Splenectomy may alleviate hypersplenism Enzyme replacement therapy is available, but is expensive Acute infantile form – splenomegaly, neurological degeneration with seizures Carrier detection and prenatal diagnosis are possible

mucopolysaccharidoses

☒ Progressive multisystem disorders which may affect the neurological, ocular,cardiac and skeletal systems Hepatosplenomegaly is usually present.

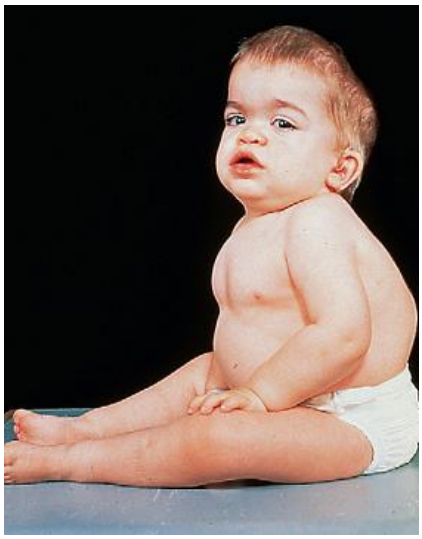
☒ **present** with developmental delay following a period Of essentially normal growth and development up to 6–12 months of age.
Children may show some loss of skills ,It is only in the second 6 months of life that the characteristic facies begin to emerge, with coarsening of the facial featuresandprominent foreheaddue to frontal bossing

☒ **diagnosis:** is made by identifying the enzyme defect and the excretion in the urine of the major storage substances, the glycosaminoglycans (GAGs).

☒ **Treatment:**

1.supportive

2.Successful enzyme replacement by bone marrow transplantation has been performed but cannot reverse any established neurological abnormality.



Eyes	Corneal clouding
	Retinal degeneration
	Glaucoma
Skin	Thickened skin
	Coarse facies
Heart	Valvular lesions
	Cardiac failure
Neurology	Developmental regression
Skeletal	Thickened skull
	Broad ribs
	Claw hand
	Thoracic kyphosis
	Lumbar lordosis
Other	Hepatosplenomegaly
	Carpal tunnel syndrome
	Conductive deafness
	Umbilical and inguinal hernias

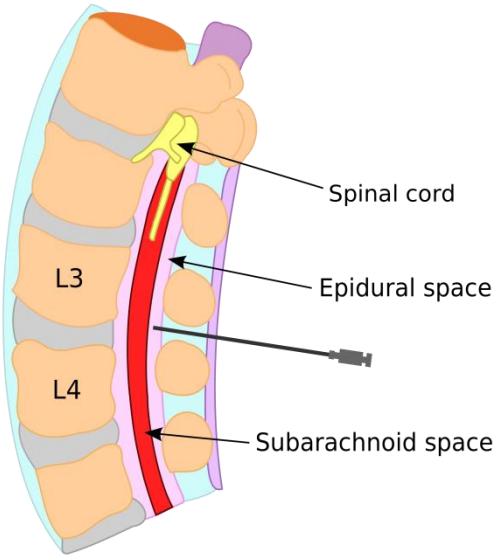
Type	Inheritance	Cornea	Heart	Brain	Skeletal
MPS I (Hurler)	AR	+++	++	+++	++
MPS II (Hunter)	X-linked	–	+	++	+
MPS III (Sanfilippo)	AR	±	–	+	+
MPS IV (Morquio)	AR	+	+	–	+++
MPS VI (Maroteaux–Lamy)	AR	+++	++	–	++

pseudotumor cerebri

☒ neurological disorder that is characterized by increased intracranial pressure (pressure around the brain) in the absence of a tumor or other diseases

CAUSES:

1. Idiopathic
2. Some risk factors:
3. high-dose vitamin A derivatives (e.g. isotretinoin for acne)
4. long-term tetracycline antibiotics (for a variety of skin conditions)
5. hormonal contraceptives.
6. Corticosteroid
7. Nalodoxic
8. Nitrofurination
9. Hyperthyroidism
10. Hypoparathyroidism
11. Thrombosis EX: venous sinus thrombosis



FEATURES OF INCREASED ICP

Before fontanel closure:

1. Tense, bulging anterior fontanel
2. Irritable and poor feeding

After closure of fontanels:

1. Headache, (irritability) (92–94%) → worse in the morning, generalized Throbbing innature. nausea and vomiting.
   Pulsatile tinnitus→ whooshing sensation in one or both ears (64–87%) Severe
2. Blurred of vision
3. Projectile vomiting (in the morning, not preceded by nausea)
4. Cushing response (hypertension & bradycardia)

MANGMENT:

1. Remove risk factor
2. Wt loss
3. Lasix-acetazolamide
4. Spinal tap (not used if ant fontanella opened)
5. Shunt

لاتنسونا من صالح الدعاء
بالتوفيق للجميع
د.علي بالخير